

REVIEW ARTICLE

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# A systematic review of the efficacy of cochlear implantation in adults with auditory neuropathy spectrum disorders

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## Abstract

**Background** Auditory neuropathy spectrum disorder (ANSO) is characterized by normal outer hair cell functioning alongside compromised auditory nerve activity. Our study is designed with the aim of evaluating the effectiveness of cochlear implantation in addressing this disorder among adult individuals.

**Methods** The systematic review was carried out using PRISMA guidelines. The review resulted in 288 articles related to the topic. Among these, 11 articles met the inclusion and exclusion criteria that were included for the study. The studies were evaluated using the Revised Tool for the Quality Assessment of Diagnostic Accuracy Studies.

**Results** We conducted a comprehensive review by examining articles sourced from various databases to investigate the effectiveness of cochlear implantation in individuals diagnosed with auditory neuropathy spectrum disorder (ANSO). The majority of the studies indicate positive outcomes associated with cochlear implantation, resulting in improved audibility and enhanced speech perception abilities. Nonetheless, the prognosis is influenced by a range of factors including the specific location of the auditory lesion, the underlying cause of ANSO, and the nature of post-operative training. The evaluation of article quality revealed a minimal risk of bias, indicating a robust foundation for the conclusions drawn.

**Conclusion** The review substantiates the effectiveness of cochlear implantation in addressing the needs of adult individuals dealing with auditory neuropathy spectrum disorder (ANSO). Notably, the review emphasizes that the decision to recommend a cochlear implant should be made in light of the insights derived from genetic testing. However, it is important to highlight that the existing literature lacks a sufficient number of experimental studies featuring appropriately sized samples, underscoring the necessity for future research endeavors to bridge this gap and enhance our understanding of this intervention's potential.

**Keywords** Auditory neuropathy, Synaptopathy, Cochlear implant, Adult, Efficacy

## Background

Auditory neuropathy spectrum disorder (ANSO) is identified by intact outer hair function but compromised auditory nerve activity [1]. The underlying cause of ANSO is a disturbance in the coordinated neuronal

activity of the auditory nerve, and the site of the impairment can be categorized as either pre-synaptic or post-synaptic [2]. Pathological disruptions before the synapse can manifest in the inner hair cells or at the junction between inner hair cells and type 1 afferent nerve fibers. Conversely, a post-synaptic lesion refers to an anomaly within the auditory nerve itself [3].

The prevalence of ANSO diagnosis is on the rise with each passing year [4]. Although the exact prevalence of ANSO remains uncertain, estimates range from less than

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1 to 10% among individuals with hearing impairment [4]. ANSD is linked to a range of causative factors including hyperbilirubinemia and anoxia, as well as genetic and syndromic conditions like Charcot-Marie-Tooth syndrome, along with infectious processes [1]. Research involving animal models has identified diverse locations of dysfunction, including inner hair cells, synapses, and auditory nerve fibers, shedding light on the genetic aspects [5]. The terms “auditory neuropathy” and “auditory synaptopathy” have emerged to delineate ANSD due to the impaired IHC ribbon synapse functionality and neural fiber dysfunction [3].

The behavioral audiological characteristics of individuals with ANSD can exhibit remarkable variability due to the fluctuating nature of their hearing loss, spanning from normal to profound levels [6]. This disorder presents diverse functional implications, where difficulties in speech perception often outweigh the extent of hearing impairment [7]. Patients with auditory neuropathy have access to a range of treatment possibilities. Over the years, clinicians have explored various therapeutic approaches such as auditory training, fitting of hearing aids, utilization of assistive listening devices, and even cochlear implantation, tailored to the severity of the issues [8]. However, it is important to note that none of these methods has achieved complete efficacy. Among the rehabilitation options for adults with auditory neuropathy, Cochlear implantation stands out as a widely employed choice [4]. This can be attributed to the fact that implanted electrodes directly transmit electrical impulses to the auditory nerve, bypassing the presynaptic inner hair cells and their associated synapses that are involved in the auditory nerve’s firing process [9].

### Main text

The literature has demonstrated the advantages of cochlear implantation for both children and adults afflicted with auditory neuropathy spectrum disorders [7, 8]. Cochlear implants have proven successful in effectively stimulating the auditory system, leading to substantial perceptual improvements in individuals of varying ages with auditory neuropathy. However, despite the available evidence, the outcomes of cochlear implantation remain somewhat ambiguous. It is important to note that not all children and adults with ANSD experience benefit from cochlear implantation. Several factors come into play, including residual hearing, socioeconomic circumstances, and the duration of auditory deprivation, all of which exert an influence on the outcomes of cochlear implantation. The primary determinant of these outcomes is the location of the auditory pathway lesion [3].

In accordance with Shearer et al.’s research, the genetic site of the lesion and its impact on spiral ganglion

function can serve as predictive indicators of cochlear implant outcomes in auditory neuropathy spectrum conditions [3]. Their study revealed that individuals with mutations in genes responsible for spiral ganglion dysfunction (such as OPA1, DFNB59, AIFM1, DIAPH3) exhibited notably poorer speech perception outcomes compared to those with mutations in genes associated with presynaptic and postsynaptic cochlear dysfunction (such as OTOF, SLC17A8, CACNA1D, CABP2) [3].

Cochlear implants (CIs) have emerged as the most effective rehabilitation method for enhancing auditory capability and communication skills in patients grappling with auditory neuropathy [10]. A review conducted by Roush et al. [11] underscored the benefits of cochlear implantation for children afflicted by ANSD. However, there is a noticeable dearth of comprehensive review studies that establish the effectiveness of CIs for adults with ANSD. In many cases, adults who experience post-lingual auditory neuropathy often choose against cochlear implantation for rehabilitation due to various constraints including financial costs, potential surgical complications, and the lack of clear postoperative prognostic evidence associated with cochlear implants. Regrettably, there is a limited number of experimental design studies that shed light on the outcomes of cochlear implantation for adults with auditory neuropathy. The speech perception results following cochlear implantation display variability among individuals [12].

Dean et al.’s investigation revealed positive advancements in speech perception among both low and high-performing groups of individuals with ANSD who underwent cochlear implantation. Their study also highlighted a significant correlation between pre-implant pure tone averages and improved speech performance post-implantation [7]. Similarly, the study done by Yüksel and Çiprut [13] demonstrated that ANSD patients who received cochlear implants could derive music-related and psychoacoustic benefits similar to those observed in patients with sensorineural hearing loss (SNHL). Nonetheless, there remains a scarcity of comprehensive literature reviews that combine all available information cohesively, offering a more distinct comprehension of the efficacy of cochlear implantation for adults with ANSD.

Our research seeks to conduct a comprehensive systematic review encompassing all pertinent information in adults with auditory neuropathy spectrum disorder (ANSD). The overarching goal of this review is to provide a clearer understanding of the effectiveness of cochlear implants in this particular demographic. This study aims to address the following research inquiries:

1. To what extent is cochlear implantation efficacious for adults diagnosed with ANSD?

2. What are the key prognostic indicators that can shed light on the outcomes of cochlear implantation in this population?
3. What are the pertinent factors that need to be taken into account when considering the recommendation of cochlear implants for the adult segment of the ANSD population?

By systematically assessing and synthesizing existing literature, this review seeks to offer a comprehensive overview that can contribute to a more informed understanding of the potential benefits, prognostic markers, and relevant considerations pertaining to the implementation of cochlear implants in adults with ANSD.

## Methods

The research was carried out in adherence to the ethical principles set forth by the institutional review board. Prior to commencing the study procedures, registration was completed on PROSPERO (Registration ID: CRD42021262786), and this registration information is available as [supplemental material](#). The process of literature exploration was conducted in strict accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) criteria [14]. The assessment of study quality was undertaken using the Revised Tool for the Quality Assessment of Diagnostic Accuracy Studies [15].

## Search engines

Various databases were employed during the search process, including Google Scholar, PubMed, Comdisdome, Web of Science, Schematic Scholar, and Elsevier. A comprehensive database search was executed using pertinent keywords related to the subject matter. These keywords encompassed terms such as “cochlear implant,” “ANSD,” “Auditory neuropathy,” “Auditory dys-synchrony,” and “Adult,” and their respective variations were incorporated using appropriate Boolean operators. Articles were meticulously selected based on predefined inclusion and exclusion criteria.

Specifically, the study encompassed peer-reviewed articles available until the year 2021, focusing on cochlear implantation and ANSD within the adult population. The articles had to be written in English and involve human subjects. To ensure the integrity of the study, duplicate studies were excluded from the primary sample. Articles centered on cochlear implantation in pediatric cases of ANSD, those employing animal models, and articles not in English were excluded. Similarly, review articles, investigations in histopathology, and studies utilizing duplicated data were also omitted from consideration in this study.

## Data extraction (selection and coding)

The review process adhered to the PRISMA guidelines established by Moher et al. [14]. To identify pertinent articles, an extensive compilation of keywords was utilized. The articles underwent a selection process based on predefined inclusion and exclusion criteria. This involved an initial assessment of titles followed by abstract screening. Full texts of all eligible articles were thoroughly assessed in alignment with the predetermined criteria to determine their eligibility.

The search and screening procedures were conducted by three reviewers, and any disparities that arose during the screening phase were addressed through collaborative discussions. Information from the selected studies was extracted using a pre-designed form, which had undergone preliminary testing (pre-piloted). This rigorous approach ensures the systematic and robust evaluation of the literature, aligning with the established guidelines for transparent and thorough research synthesis.

## Results

The outcomes derived from the review were consolidated through a structured approach that encompassed multiple aspects. Initially, the process involved the extraction of essential information from the chosen studies. This encompassed pertinent details and findings from each study that were of relevance to the subject at hand. Additionally, a meticulous quality analysis was performed on the selected articles. This evaluation aimed to assess the methodological accuracy, robustness of data collection, and overall rigor of each study. The intention was to ascertain the reliability and validity of the evidence presented within the articles. Lastly, a comprehensive summary of the selected articles was formulated. This summary encompassed a synthesis of the accumulated evidence regarding the efficacy of cochlear implantation in the context of auditory neuropathy in adults (ANSD). By consolidating the findings, this summary aimed to provide a clear and concise overview of the benefits observed in adults with ANSD who underwent cochlear implantation. This structured approach ensured that the results were systematically compiled, analyzed for quality, and succinctly presented, enhancing the overall comprehensibility and reliability of the conclusions drawn from the review.

## Article selection

The initial phase of the search process, alongside the application of predefined inclusion and exclusion criteria, resulted in the identification of 11 papers that were subsequently subjected to quality analysis and synthesis. A comprehensive database search initially yielded

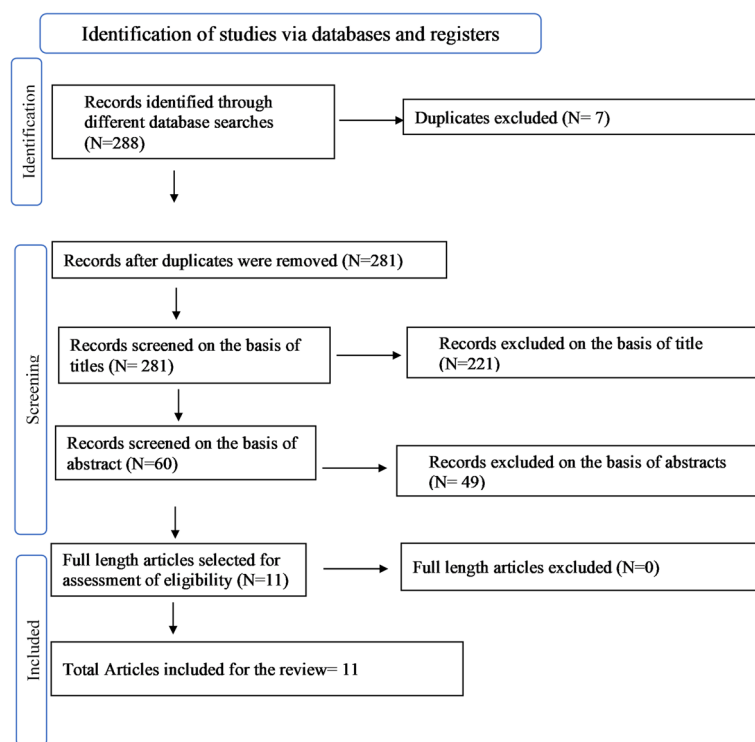
a total of 288 articles. However, 7 of these articles were excluded due to duplication, resulting in 281 articles for title screening. Through this process, 221 papers were excluded based on their titles. Subsequently, abstract screening further excluded 49 articles. Ultimately, 11 articles were selected for a thorough examination that satisfied the stipulated selection criteria, and these 11 articles formed the foundation of the review. To ensure the robustness of the selection process, an inter-judge selection methodology was employed, and any discrepancies that arose were resolved through collaborative discussions. The selected articles encompassed a range of research designs, including experimental, retrospective, and case series analyses, providing a diverse scope for the review. A graphical representation of this information can be found in the PRISMA table, as depicted in Fig. 1. This comprehensive overview outlines the progression of the selection process and highlights the rigorous approach undertaken in the systematic review.

**Quality analysis of the selected articles**

In order to comprehend nonrandomized studies effectively, a comprehensive quality assessment is imperative. To achieve this, the study’s quality and robustness were evaluated using Whiting et al.’s QUADAS-2 tool [15]. This standardized tool has been designed specifically to gauge the quality and precision of diagnostic studies,

making it an appropriate choice for systematic reviews. The QUADAS-2 tool is organized into four key domains: patient selection, index test, reference standard, and flow and timing. This structure enables a comprehensive evaluation of potential biases and applicability concerns in diagnostic accuracy studies. Each aspect within the QUADAS-2 tool was evaluated using a response classification of “yes,” “no,” or “unclear.” This assessment was done to identify the potential risk of bias, sources of variability, and overall study quality. By employing this structured approach, the review aimed to ensure a robust evaluation of the chosen studies, thereby enhancing the reliability and validity of the conclusions drawn from the synthesized evidence.

The majority of the studies selected for evaluation demonstrated the use of well-calibrated instrumentation, standardized questionnaires, and appropriate methodologies. Moreover, a substantial portion of these studies exhibited robust control measures, with careful case and control selection processes that ensured a representative sample. While a few studies displayed a high risk of bias, the majority maintained effective control over research parameters, resulting in a low risk of bias. However, a common limitation observed across most studies was the inability to account for potential confounding factors, which could potentially introduce bias into the results. Despite this limitation, the studies generally exhibited



**Fig. 1** Flowchart depicting the selection process of the articles in the systematic review

minimal risk of bias and source of variance in terms of patient selection, flow, timing, and index test. The comprehensive overview of the quality assessment outcomes is illustrated in Table 1, which provides a clear summary of the quality analysis conducted on the selected studies. This systematic evaluation contributes to the overall reliability and credibility of the synthesized evidence in the review.

**Summary of the data extraction**

The data extraction process involved systematically gathering and categorizing information from the selected papers, encompassing several key criteria: author and year of publication, research design, research question, population type, techniques employed, outcomes, and discussions. All of the included research studies were published up to 2021 and primarily consisted of experimental studies, retrospective studies, case reports, case series, and cohort designs. The participants in these studies were exclusively adults who had received a diagnosis of ANSD and subsequently underwent cochlear implantation.

Throughout the selected studies, a variety of preoperative, intraoperative, and postoperative tests were conducted to assess the effectiveness of cochlear implantation. In the preoperative phase, the most commonly employed testing parameters included behavioral tests, electrophysiological tests, and radiological imaging.

Behavioral tests encompassed pure tone audiometry, speech audiometry, and speech perception in noise. Electrophysiological assessments included auditory brainstem response (ABR), otoacoustic emissions (OAE), and electrocochleography (ECoCHG). Radiological imaging was done using CT scans and MRI scans. During the intraoperative phase, commonly used tests involved electrical compound action potential (ECAP), electrical stapedial reflex threshold (ESRT), and neural response telemetry (NRT). The postoperative comparison was centered on evaluating improvements in various domains, including speech perception, speech production, music perception, spectral perception, and temporal perception. This comparison aimed to explain the extent of enhancement brought about by cochlear implantation, by contrasting pre-operative and post-operative outcomes in these domains.

**Postoperative findings reported across the studies**

Throughout the range of studies reviewed, diverse outcomes have been observed. Positive effects have been documented using both behavioral and electrophysiological metrics. Within the realm of behavioral measures, benefits have been demonstrated in terms of improved audibility, enhanced speech perception outcomes, and heightened music perception capabilities. Conversely, electrophysiological assessments have indicated benefits through enhancements in neural response telemetry

**Table 1** Tabular representation of quality analysis of the studies selected for the review using QUADAS-2

Study Authors	Risk of Bias				Applicability concern		
	Patient Selection	Index test	Reference standard	Flow and timing	Patient selection	Index test	Reference standard
Dutt et al.(2021)	L	L	L	L	H	L	L
Mason et al. (2003)	L	L	L	L	H	L	L
Ji et al.(2014)	L	L	L	L	L	L	L
Santarelli et al. (2015)	L	L	L	L	?U	L	L
Dean et al.(2013)	L	L	L	L	L	L	L
Yüksel & Çıprut.(2020)	L	L	L	L	L	L	L
Postelmans & Stokroos. (2006)	L	L	L	L	L	L	L
Derinsu et al. (2007)	L	L	L	L	L	L	L
Leenheer et al.(2008)	L	L	L	L	L	L	L
Matsuda & Kaga.(2021)	H	L	L	L	L	L	L
Katada et al. (2005)	H	L	L	L	?U	L	L





and the electrical auditory brainstem response (eABR) response. These findings collectively underscore the multifaceted nature of the benefits brought about by cochlear implantation for adults with ANSD, encompassing both perceptual and physiological enhancements.

Mason et al.'s study [16] highlighted a crucial factor in post-operative outcomes: the presence of a response during promontory stimulation. Their findings demonstrated that individuals who exhibited improvement in subjective measures like speech perception and sound quality post-cochlear implantation were those who displayed a positive response during promontory testing. They went further to suggest that patients who did not demonstrate auditory perception benefits during promontory testing might not derive advantages from implantation, indicating the importance of this criterion in the decision-making process. They also reported avoiding implantation in patients who do not demonstrate benefit on auditory perception during promontory testing. Similarly, Dutt et al. [17] reported similar conclusions in terms of subjective measures, reporting beneficial outcomes. However, unlike Mason et al.'s study, Dutt et al. did not find corresponding benefits in objective measures. This discrepancy underscores the complex interplay between subjective and objective measures in evaluating the effectiveness of cochlear implantation for individuals with ANSD. These studies collectively contribute to a deeper understanding of the subtle outcomes associated with cochlear implantation in this population.

In line with the aforementioned findings, Ji et al. [18] contributed to the understanding of postoperative performance prediction. They found that the absence of preoperative and postoperative electrical compound action potential (ECAP) did not preclude a substantial improvement in patient performance after cochlear implantation. This observation led them to infer that cochlear implants have the potential to restore the synchronization of the auditory nerve pathway for a significant portion of patients with auditory neuropathy, irrespective of the severity of their hearing loss. As a result, cochlear implants could be recommended as a preferred intervention for individuals with auditory neuropathy who do not experience benefits from conventional hearing aids. This underscores the potential efficacy of cochlear implants in addressing the challenges associated with auditory neuropathy spectrum disorder. Indeed, the utilization of electrically evoked auditory brainstem response (eABR) as an intraoperative measure for assessing postoperative performance has been a consistent approach across various studies. The collective findings from these studies underscore a noteworthy observation: the presence of eABR responses does not serve as an absolute predictor of the ultimate prognosis with a cochlear implant. Contrary to

the common notion, certain cases within the spectrum of ANSD have demonstrated comparable performance outcomes in adults with absent eABR responses, paralleling the outcomes in adults with present eABR responses.

This intricate interplay between eABR responses and post-operative outcomes highlights the heterogeneous nature of auditory neuropathy and the potential for individuals with absent eABR responses to still achieve positive results through cochlear implantation. Despite this, it's important to note that eABR results could serve as valuable prognostic indicators for future performance. This recognition showcases the complexity of the relationship between physiological measures and functional outcomes in the context of cochlear implantation for adults with ANSD.

The majority of the studies included in the review have underscored the favorable outcomes associated with cochlear implantation for adult individuals with auditory neuropathy spectrum disorder (ANSD). However, the magnitude of improvement is dependent upon several factors, including the site of the lesion, the individual's preoperative speech recognition performance, and the efficacy of postoperative training. Interestingly, a study highlighted that the success of cochlear implantation in ANSD depends on the location of the lesion [19]. In cases where the lesion is situated within the auditory nerve itself, the cochlear implant might not effectively transmit information to higher auditory structures. On the other hand, when the lesion involves inner hair cells or the synapse with afferent nerve fibers, the likelihood of benefiting from cochlear implantation is notably higher.

While the benefits of cochlear implantation in ANSD are influenced by etiology and pathophysiology, it remains the most effective rehabilitation strategy currently available for individuals who do not experience improvements from conventional amplification approaches. The significance of proper post-surgery rehabilitation training cannot be underestimated, as it plays a pivotal role in achieving positive outcomes. A comprehensive overview of the 11 studies included in the review can be found in the [supplementary table](#), encapsulating the key findings and insights collected from the collective research endeavors.

## Discussion

This systematic review aims to assess the effectiveness of Cochlear implantation (CI) in the context of adults diagnosed with auditory neuropathy spectrum disorder (ANSD). Cochlear implantation has proven to be an efficacious rehabilitative intervention for adults exhibiting severe to profound cochlear hearing impairment [20, 21]. Notably, Cochlear implantation has also been employed as a rehabilitative avenue for individuals with neural

hearing impairments such as ANSD [22]. Nevertheless, a notable dearth of comprehensive empirical evidence exists concerning the effectiveness of CI in this specific context. Consequently, the primary objective of this investigation is to bridge this existing knowledge gap by precisely reviewing the relevant literature.

Certainly, acquiring a cochlear implant to address serious hearing difficulties can come with a significant cost. Before recommending this treatment widely, especially given its high expense, it is crucial to understand its efficacy. If people do not experience substantial improvement after undergoing the treatment, it can create challenges for them and their families. As a result, our study aimed to investigate the effectiveness of cochlear implants for individuals with auditory neuropathy spectrum disorder (ANSD). Notably, this study is the first of its kind to examine the effectiveness of cochlear implants in adults dealing with ANSD.

The comprehensive analysis we conducted revealed a favorable outcome associated with the utilization of cochlear implants (CI) among adults with auditory neuropathy spectrum disorder (ANSD). Comparable positive outcomes have been documented by Roush et al. [11] and Humphriss et al. [23] in the context of children. It is important to note that only a small number of individuals, particularly those who choose cochlear implants after language development in adulthood, participate in the studies. Consequently, there is a deficiency in experimental studies with adequately sized samples within the existing literature. To mitigate this limitation, our review encompassed not only research articles but also case reports and case series, which might have influenced the assessment of article quality. In total, our review encompassed 11 articles, collectively indicating that individuals with ANSD experience benefit from cochlear implantation, as evident from either behavioral evaluations or electrophysiological measurements [17].

#### **Behavioral measure outcome after cochlear implantation**

The outcomes of our review revealed that following cochlear implantation, individuals with auditory neuropathy spectrum disorder (ANSD) reported experiencing subjective advantages such as heightened audibility and improved speech comprehension. Various subjective audiological assessments, including pure tone audiometry (PTA), speech-in-noise test (SPIN), and speech recognition testing, have been used in the literature to ascertain the benefits of cochlear implantation. Although those with ANSD tend to struggle more with difficulties in speech recognition rather than audibility issues, patients reported a subjective improvement in audibility subsequent to cochlear implantation [24].

In relation to speech recognition scores, conflicting results are present in the literature concerning the benefits. Some studies did not document any advantages of cochlear implantation, both in quiet and noisy environments [16]. Conversely, a subset of studies did report benefits, demonstrating enhanced performance in speech recognition scores following cochlear implantation [25, 26]. This disparity could be attributed to variations in the specific locations of the auditory lesion within the distinct participant groups recruited for the studies. Consequently, these findings suggest that conducting genetic testing for individuals with ANSD is crucial to pinpoint the exact site of the auditory lesion prior to considering cochlear implantation as an option. The decision to recommend a cochlear implant should be informed by the results of genetic testing.

#### **Electrophysiological measure outcome after the cochlear implantation**

Various objective measures, such as eABR (electrically evoked auditory brainstem response), eCAP (electrically evoked compound action potential), and ECochG (electrocochleography), have been utilized in the literature to assess the definite advantages of cochlear implantation. Researchers aimed to demonstrate the benefits that occur after the surgical procedure by assessing compound action potentials subsequent to implantation. Some studies indicated the absence of eCAP in ANSD individuals who received cochlear implants [18]. Conversely, a different set of studies showcased an enhancement in the eABR response, manifested by the appearance of the fifth peak post-implantation in ANSD patients, even when eCAP was absent [25]. These findings underline the influence of multiple factors, including the root cause of ANSD and the specific location of the auditory lesion, on the extent of improvement achievable through cochlear implantation. Similarly, investigations have also identified advantages concerning music perception through psychoacoustic assessments in individuals with ANSD [10, 13]. As a result, prior to recommending cochlear implantation for adults with ANSD, medical practitioners must consider various factors that could impact the efficacy of cochlear implantation.

#### **Implications of the study**

Through our investigation on the efficacy of cochlear implantation (CI), the outcomes of our study provide valuable insights to clinicians specializing in the field of cochlear implantation. Specifically, our findings can aid clinicians in determining the appropriate criteria for selecting candidates for this intervention. Furthermore, our study underscores the diverse array of factors that exert an influence on the outcomes following the

surgical procedure. This, in turn, facilitates informed decision-making regarding the recommendation of cochlear implants for adult individuals with auditory neuropathy spectrum disorder (ANSD). Additionally, our review sheds light on the heterogeneous aspects of assessment that warrant attention before recommending cochlear implantation as a viable option for individuals affected by ANSD. This comprehensive approach to assessment is crucial for ensuring the best possible outcomes for patients in need of such interventions.

### Conclusion and limitation of the study

The review presents compelling evidence that supports the effectiveness of cochlear implantation among adult individuals with auditory neuropathy spectrum disorder (ANSD). It is important to note that there is a dearth of experimental studies featuring sufficiently large sample sizes and the studies focusing on the etiology behind the disorder within the existing body of literature underscoring the necessity for future research endeavors in this area. Nonetheless, the absence of robust evidence should not act as a deterrent to the utilization of cochlear implantation within these populations. Clinicians are advised to possess a comprehensive understanding of the various factors that might impact the prognosis associated with cochlear implantation, and they should make recommendations accordingly. While acknowledging the current limitations, the advancement and widespread adoption of this beneficial technology within these populations on a global scale necessitates the accumulation of stronger evidence.

### Supplementary Information

The online version contains supplementary material available at <https://doi.org/10.1186/s43163-023-00542-9>.

**Additional file 1: Table S1.** Study characteristics of reviewed articles.  
**Table S2.** Summary of the reviewed articles.

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### Authors' contributions

SA was involved in the concept development, study design, stimulus preparation, analysis of the results, interpretation, and writing the manuscript; YS was involved in concept development, study design, stimulus preparation, analysis of the results, and writing the manuscript; PP was involved in the concept development and study design, stimulus preparation, and writing the manuscript.

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### Availability of data and materials

The datasets analyzed during the current study are available from the corresponding author upon reasonable request.

### Declarations

#### Ethics approval and consent to participate

The research followed ethical guidelines by the All India Institute of Speech and Hearing's review board, obtaining informed consent from all participants.

#### Consent for publication

Not applicable.

#### Competing interests

The author declares that they have no competing interests.

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