

CORTICAL AUDITORY EVOKED POTENTIALS IN TWO PRELINGUALLY DEAFENED ADULTS

Aju Abraham, Hari Prakash, Bellur Rajashekhar, Krishna Yerraguntla

Department of Speech and Hearing, School of Allied Health Sciences, Manipal University, Manipal, India

Corresponding author: Aju Abraham, Department of Speech and Hearing, School of Allied Health Sciences, Manipal University, Manipal, India, e-mail: ajubaslp@gmail.com

Abstract

Background: It has been reported that in prelingually deafened adults there is maturation of the late latency response (LLR) after rehabilitation using cochlear implants. Other evidence suggests that the auditory system, like any other sensory system, receives multimodal stimulation, a factor which could help the auditory system mature even in the absence of any auditory input. The current study was done on two prelingually deaf adults who had not received rehabilitation, and their latencies were within the normal mature range, suggesting their auditory cortex responses were maturely developed prior to rehabilitation.

Material and methods: Two male participants took part in the study: one was 21 years old and the other was 36, and both had congenital profound hearing loss. Using pure tone thresholds obtained earlier, a strong class hearing aid (Siemens Infronti Pro SP) was programmed using NOAH software and fitted to the right ear using the NALNL-1 fitting formula. As an objective measure of verification of hearing aid effect, auditory P1, N1, P2, and N2 were recorded using the free-field facility available in the IHS SmartEP system: a 1000 Hz pure tone was presented at 70 dB SPL (30 dB SL) and 50 dB SPL (10 dB SL) from a loudspeaker at a distance of 1 m and 45° angle.

Results: The latencies of P1, N1, P2, and N2 for both participants were recorded for 70 and 90 dB HL; all latencies were well within normal limits.

Conclusions: The results indicate that, with adequate amplification, a mature response from the auditory cortex can be obtained even in adults who have profound hearing loss and who have been deprived of auditory stimuli since a prelingual age.

Keywords: age factors • correction of hearing impairment • evoked potentials • auditory

LOS POTENCIALES AUDITIVOS EVOCADOS DE CORTEZA AUDITIVA EN LOS ESTADOS DE UNA LATENCIA ALTA EN DOS ADULTOS CON SORDERA PRELINGUAL

Resumen

Introducción: Tras la rehabilitación asociada al uso de los implantes cocleares, en adultos con la sordera prelingual, se observa el desarrollo de una respuesta auditiva de latencia tardía (LLR – late latency response). Otras pruebas sugieren que el sistema auditivo, como cualquier otro sistema de órganos sensoriales, recibe la estimulación multimodal- un factor que podría ayudar al sistema auditivo en su desarrollo incluso en el caso de la ausencia de la información auditiva. El estudio ha sido realizado en dos adultos con la sordera prelingual, que no habían sido rehabilitados, y la latencia aparecía en el rango normal para su edad, lo que indica que las respuestas auditivas de corteza auditiva estaban bien desarrolladas antes de la rehabilitación.

Materiales y métodos: En el estudio participaron dos hombres, de 21 y de 36 años. Los dos tenían una profunda pérdida auditiva congénita. Utilizando el estudio anterior de los umbrales auditivos, se ajustaron los audífonos de alta potencia (Siemens Infronti Pro SP) mediante el software NOAH. El dispositivo se colocó en el oído derecho utilizando la fórmula NAL NL-1. Como una forma objetiva para verificar el efecto del funcionamiento del audífono, se ha hecho la grabación de los potenciales P1, N1, P2 y N2 en las condiciones del campo abierto en el sistema IHS SmartEP: sonido de la frecuencia de 1000 Hz fue transmitido con la intensidad de 70 dB (30 dB SL) y 50 dB SPL (10 dB SL) a través de un altavoz colocado a 1 metro y con el ángulo de 45°.

Resultados: Las latencias de los potenciales P1, N1, P2 y N2 para ambos participantes del estudio fueron registrados para 70 y 90 dB HL; todas las latencias han salido dentro de la norma.

Conclusiones: Los resultados muestran que con un refuerzo adecuado, las respuestas desarrolladas de la corteza auditiva pueden aparecer incluso en adultos con una pérdida auditiva profunda, que no presentan estímulos auditivos desde la etapa prelingual.

Palabras clave: factores de edad • corrección de la pérdida auditiva • potenciales auditivos evocados

СЛУХОВЫЕ ВЫЗВАННЫЕ ПОТЕНЦИАЛЫ ОТ СЛУХОВОЙ КОРЫ ПРИ ВЫСОКОЙ ЛАТЕНТНОСТИ У ДВУХ ВЗРОСЛЫХ С ПРЕЛИНГВАЛЬНОЙ ТУГОУХОСТЬЮ

Изложение

Введение: У взрослых лиц с прелингвальной глухотой после реабилитации, связанной с использованием улиточных имплантатов, развивается длиннотентный ответ (LLR – *late latency response*). Другие данные свидетельствуют о том, что слуховая система, как и любая другая система органов чувств, получает мультимодальную стимуляцию – фактор, который может способствовать развитию слуховой системы даже в случае отсутствия слуховой информации. Исследование было проведено на двух лицах с прелингвальной глухотой, которые не были реабилитированы, а латентность появилась в нормальном для их возраста объёме, что указывает на то, что слуховые ответы от слуховой коры были хорошо развиты до реабилитации.

Материал и методы: В исследовании приняли участие двое мужчин в возрасте 21 и 36 лет. Оба имели врождённую глубокую тугоухость. С учётом результатов проведённого ранее исследования порогов слышимости, был подобран слуховой аппарат высокой мощности (Siemens Infiniti Pro SP) с помощью программного обеспечения NOAH. Он был размещён в правом ухе при помощи формулы NAL NL-1. В качестве объективного способа верификации эффекта действия слухового аппарата была проведена запись потенциалов P1, N1, P2 и N2 в условиях свободного поля в системе IHS SmartEP: звук частотой 1000 Гц подавался с интенсивностью 70 дБ (30 дБ SL) и 50 дБ SPL (10 дБ SL) через динамик, размещённый на расстоянии 1 м под углом 45°.

Результаты: Латентности потенциалов P1, N1, P2 и N2 для обоих участников были зарегистрированы для 70 и 90 дБ HL; все латентности были в норме.

Выводы: Результаты показывают, что при соответствующем усилении развитые ответы от слуховой коры могут появиться даже у взрослых с глубокой тугоухостью, у которых отсутствуют слуховые импульсы с прелингвального периода.

Ключевые слова: возрастные факторы • коррекция тугоухости • слуховые вызванные потенциалы

ŚŁUCHOWE POTENCJAŁY WYWOŁANE Z KORY ŚŁUCHOWEJ U DWÓCH OSÓB DOROSŁYCH Z GŁUCHOTĄ PRELINGWAŁNĄ

Streszczenie

Wprowadzenie: U osób dorosłych z głuchotą prelingwalną późna odpowiedź latencyjna (LLR – *late latency response*) rozwija się po rehabilitacji związanej z używaniem implantów ślimakowych. Inne dowody wskazują na to, że system słuchowy, jak każdy inny system narządów zmysłu, odbiera stymulację wielomodalną – czynnik, który mógłby pomóc systemowi słuchowemu rozwijać się nawet w przypadku braku informacji słuchowej. Badanie zostało przeprowadzone na dwóch osobach z głuchotą prelingwalną, które nie były rehabilitowane, a latencja pojawiła się w zakresie normalnym dla ich wieku, co wskazuje, że słuchowe odpowiedzi z kory słuchowej były dobrze rozwinięte przed rehabilitacją.

Material i metody: W badaniu wzięło udział dwóch mężczyzn w wieku 21 i 36 lat. Obaj mieli wrodzony głęboki niedosłuch. Korzystając z wcześniejszego badania progów słyszenia, dopasowano aparat słuchowy dużej mocy (Siemens Infiniti Pro SP) za pomocą oprogramowania NOAH. Umieszczono go w prawym uchu przy pomocy formuły NAL NL-1. Jako obiektywny sposób weryfikacji efektu działania aparatu słuchowego, dokonano zapisu potencjałów P1, N1, P2 i N2 w warunkach wolnego pola w systemie IHS SmartEP: dźwięk o częstotliwości 1000 Hz został podany z natężeniem 70 dБ (30 dБ SL) i 50 dБ SPL (10 dБ SL) przez głośnik umieszczony w odległości 1 m i pod kątem 45°.

Wyniki: Latencje potencjałów P1, N1, P2 i N2 dla obu uczestników zostały zarejestrowane dla 70 i 90 dБ HL; wszystkie latencje były w normie.

Wnioski: Wyniki wskazują, że przy odpowiednim wzmocnieniu, rozwinięte odpowiedzi z kory słuchowej mogą pojawić się nawet u osób dorosłych z głębokim niedosłuchem, u których nie ma bodźców słuchowych od okresu prelingwalnego.

Słowa kluczowe: czynniki wiekowe • korekta niedosłuchu • słuchowe potencjały wywołane

Background

Various researchers have reported that CAEP latency decreases and amplitude increases as a function of age, at least until age 10, so it can be used as a tool to assess auditory maturation. Investigators tend to agree that infants' responses are dominated by a large positive peak around 200 to 300 ms, and as they get older there is a decrease in P1 latency and newer peaks emerge [1]. Studies agree that the development of the central auditory system is moulded by acoustic experience [2–6]. Since the auditory system of the congenitally deaf individual does not have any input, the shaping of the auditory system through experience does not occur and the normal adaptation to the acoustic environment does not take place [4,7].

In congenitally deaf people it is possible they may receive synaptic connections from the visual system to cortical language areas, and this may lead to suppression of projections from the auditory system due to lack of acoustic stimulation. Studies on animals have reported that a significant decrease is apparent in the mean amplitude of gross synaptic currents and of individual synaptic connections in the primary auditory cortex [8]; in congenitally deaf cats there is less synchronous activation of the cortical column [9–11]. However, there appear to be no studies in the literature which evaluate auditory cortical maturation in congenitally deafened adults, and this prompted us to test the assumptions that have been formulated based on findings in animals and children.

Case presentation

Two male participants were part of the study: case 1, 21 years old, and case 2, 36 years old, both with congenital profound hearing loss. The participants did not report any middle ear pathology or other neurological conditions. There was no history of otological complaints such as ear infections and they had never undergone ENT surgery. The participants had no speech since childhood and communicated mainly using gestures. There was no family history of hearing loss. The participants had been exposed to a brief period of hearing aid use for around a month during which they were prescribed with a strong class body

level hearing aid with V cord. However, since there was little benefit from amplification, they discontinued using it and did not use any amplification devices after that.

Investigations

A calibrated pseudo two-channel audiometer (GSI-61, Grason Stadler Inc., USA) was used to estimate hearing thresholds using a stimulus delivered to the subjects through TDH-50P earphones and a Radioear B-71 bone vibrator. A GSI Tymstar middle ear analyzer was used to evaluate middle ear function. Outer hair cell function was recorded using an Echoport ILO292 (Otodynamics Ltd, UK) using v.6 software. All tests were carried out in a sound-treated room with permissible background noise level [12].

Pure tone audiometry revealed bilateral symmetric profound hearing loss with PTA >90 dB HL. Tympanometric results revealed an A-type tympanogram along with the absence of an acoustic reflex in either ear, suggesting normal middle ear function. Distortion product otoacoustic emissions were absent in both ears for F2 frequencies between 0.5 and 6 kHz. Auditory brainstem responses showed an absent bilateral wave V peak at 90 dB nHL, confirming the diagnosis of the pure tone audiogram.

Using the pure tone thresholds obtained earlier, a strong class hearing aid (Siemens Infiniti Pro SP) was programmed using NOAH software and fitted to the right ear using the NALNL-1 fitting formula. An aided audiogram was done to estimate the pure tone thresholds after hearing aid fitting, and the thresholds were within the speech spectrum for both participants (Figures 1 and 2).

As an objective measure of hearing aid verification, auditory P1-N1-P2-N2 was recorded using the free-field facility available in the IHS SmartEP system. A 1000 Hz pure tone was presented at 70 dB SPL (30 dB SL) and 50 dB SPL (10 dB SL) from a loudspeaker at a distance of 1 m and 45° angle. Gold cup electrodes were placed between Cz–A1, while the forehead served as ground. The recorded EEG was filtered online between 1 and 30 Hz. The latency and amplitude of P1, N1, P2, and N2 obtained from both

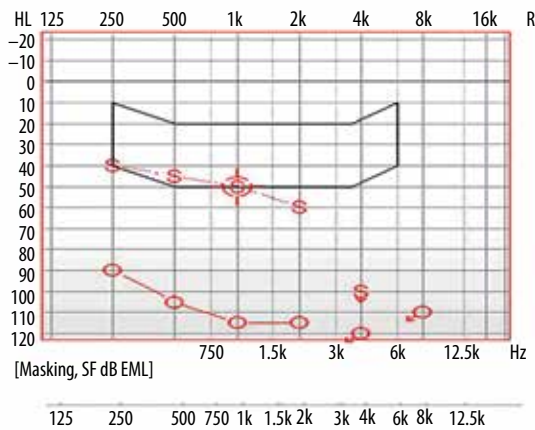


Figure 1. Unaided and aided audiogram of participant 1

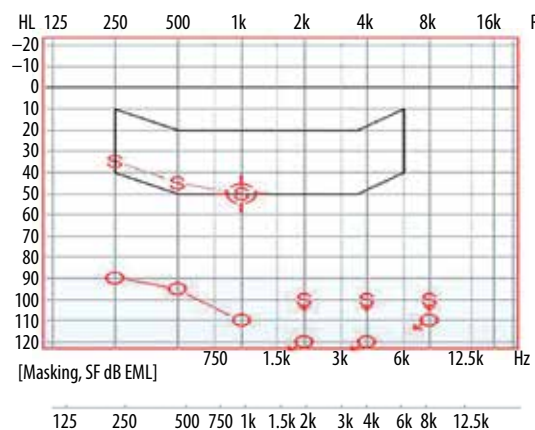


Figure 2. Unaided and aided audiogram of participant 2

Table 1. Latencies of P1, N1, P2, and N2 at 90 dB HL and 70 dB HL for two subjects

Peak	Latencies (ms) Participant 1		Latencies (ms) Participant 2	
	90 dB HL	70dB HL	90 dB HL	70dB HL
P1	98.0	79.8	81.2	84.0
N1	126.0	131.6	107.8	107.8
P2	161.0	175.0	155.4	142.8
N2	256.2	249.2	292.6	309.4

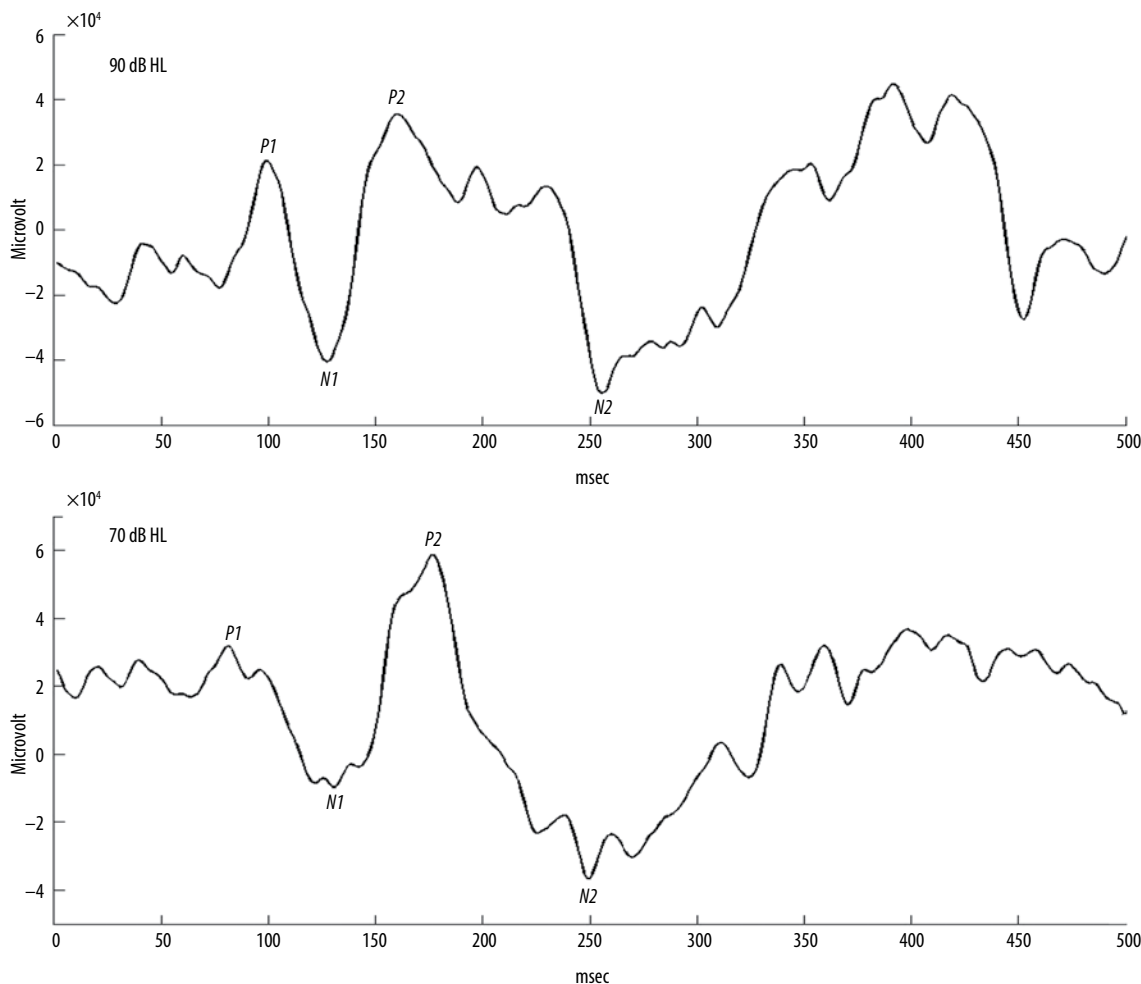


Figure 3. Aided long latency response of participant 1

participants are shown in Table 1 and in Figures 3 and 4. It is clear that CEAP components, especially P1 and N1, are within the normal latency range.

Discussion and conclusion

In the literature, changes in CEAP latency have been used as an index of auditory cortical maturation. The literature shows that lack of auditory stimulation at an early age hampers the maturation of cortical structures (as reflected by CEAPs), even if hearing aid management is provided at a later age. However, the ceiling age of this phenomenon is

unclear [1,13–16]. From our case study of two individuals, it is clearly possible to obtain a mature response from the auditory cortex (as reflected by CEAPs) even in adults who have been acoustically deprived (from a prelingual age to the time of testing).

This observation raises many questions scientifically and clinically. First, why should older children or adults acquire a normal CEAP, but not children without amplification? There are two main schools of thought regarding cortical reorganization, and one indicates that auditory deprivation leads to ‘decoupling’ of the secondary auditory cortex from

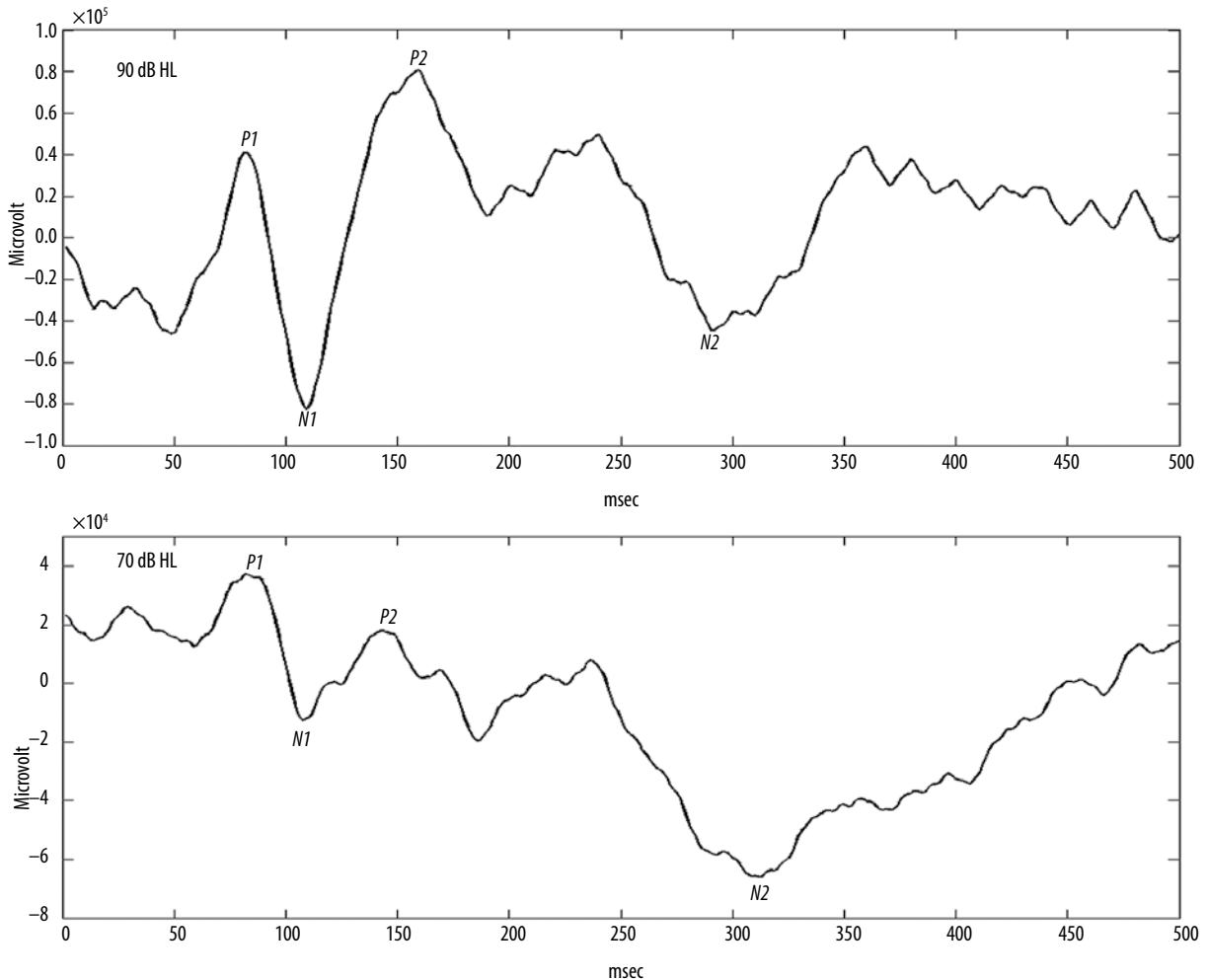


Figure 4. Aided long latency response of participant 2

the primary auditory cortex, leading to decreased cognitive modulation of higher-order auditory function [17]. Nevertheless, these studies did not comment on primary cortex reorganization, just implications about reduced processing of information in the auditory cortex due to deprivation. Indirect evidence from the somatosensory system indicates that if the cortex is not stimulated for a period, other strongly stimulated sensory regions will take over the region through the phenomenon of cross-modal reorganization [18]. Studies in the field of visual processing using fMRI and MEG also suggest that visual stimuli might activate the auditory cortex in deaf subjects [19,20].

It is possible there is something like a ‘waiting period’ during which the structures connected anatomically to other structures preserve their integrity. However, once this period is crossed (due to cortical decoupling), neighbouring structures that are already mature innervate not just the secondary cortex but possibly even primary auditory cortical neurons. Thus, when the anatomically intact connection from the central auditory pathway to the cortex is activated through amplification at a later age, there is the possibility of recording an adult-like N1 latency and amplitude, as found in our study.

Secondly, what do normal N1–P2 responses indicate in a long-deprived individual? Although the latency and morphology are normal, it might reflect the maturation of cortical neurons even when there is a lack of auditory stimulation. It is believed that a critical age 21 is important for successful rehabilitation, and that individuals receive minimal benefits after this age. However, this assumption has not been tested on older children and adults who never received any amplification. Thus, this hypothesis, mostly based on animals and the younger hearing impaired, should be tested before extrapolating it to the adult population. There is evidence from the literature which suggests there can be a mixed representation of both auditory and visual areas in the adult brain. Furthermore, recent evidence suggests that an auditory stimulus can produce evoked potentials over the visual cortex, strongly indicating that prelingually deaf adults also might show mature auditory responses due to multimodal representation in the auditory and visual cortex.

Pockett and colleagues [22] conducted a study to determine whether unimodal auditory stimuli evoke event-related potentials in brain areas normally designated as the visual cortex. The ERPs were measured from six neurologically normal adult humans and one epileptic adult human,

and the results were that unimodal auditory stimuli can evoke ERPs in the visual cortex. Burdo and colleagues [23] studied maturation of the LLR in 45 hearing-impaired individuals grouped homogeneously into five groups, one of which included prelingually deaf adults who received implants as adults. The maturation of the LLR, even at the 3rd month following cochlear implantation, showed a normal N1–P2 latency, which contradicts general findings in the

literature. This observation poses a serious limitation to using the age of the individual as a way of tracking maturational changes using cortical evoked potentials.

Clearly, there is a need for a more detailed study that includes auditory-deprived individuals of various ages to understand when and how immature responses transform to mature responses.

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