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Early Detection of Auditory Agnosia in Children during the Language Acquisition Stage : A Systematic Review of the Impact on Language Development, Cognition and Socialization

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Abstract

Background: *Pediatric auditory agnosia is a rare central auditory disorder that severely disrupts language acquisition.*

Objective: *To synthesize evidence on its etiology, diagnostic biomarkers, and clinical evolution, emphasizing the impact of early intervention.*

Methods: *A systematic review was conducted following PRISMA guidelines. Twenty-five articles (2005–2025) were selected from PubMed and Research Rabbit, focusing on children up to age 6 or longitudinal data.*

Results: *Etiologies include GRIN2A mutations, herpetic encephalitis, and epileptic encephalopathies. A hallmark finding is functional deafness despite normal peripheral hearing (ABR/DPOAE). Advanced neuroimaging (fMRI/DTI) and middle-latency responses (MLR) are robust biomarkers for mapping thalamocortical dysfunction and "non-reactive" cortical areas. Longitudinal evidence reveals that childhood agnosia may progress to permanent cortical deafness in adulthood due to retrograde degeneration of auditory radiations.*

Conclusion*: Early detection is the primary predictor of linguistic success. A multimodal diagnostic approach combining electrophysiology and functional imaging is essential. Interventions must be adaptive to address potential neural degeneration occurring decades after the initial insult.*

Keywords: Auditory agnosia; cortical deafness; Children; Language acquisition; early detection.

Introductions

Auditory perception constitutes one of the fundamental pillars of human development, playing a decisive role in language acquisition, communication, and the construction of cognitive and social competencies. The manner in which the auditory system transforms mechanical vibrations into complex neural representations is a highly specialised process, involving a sequence of anatomical and physiological mechanisms distributed across the outer, middle, and inner ear, as well as the central auditory pathways (Moore, 2012; Pickles, 2013). Understanding this trajectory is essential for recognising how specific disturbances, such as auditory agnosia, can profoundly compromise children's linguistic development. In this context, a systematic review is necessary to synthesize the available evidence, identify knowledge gaps, and suggest future directions for research and clinical practice.

Auditory System : Anatomy and Physiology

The auditory process begins in the outer ear, where the auricle, with its intricate morphology, captures and modulates incoming sound waves. The folds and depressions of the pinna selectively alter the acoustic spectrum, contributing to the spatial localisation of sounds, which is a crucial skill that enables children to focus on speech directed at them, even in noisy environments (Moore, 2012). The external auditory canal naturally amplifies frequencies between 2,000 and 5,000 Hz, precisely those that contain phonetic information relevant to language, such as vowel formant transitions and the acoustic characteristics of consonants (Pickles, 2013). Sound waves then reach the tympanic membrane, whose vibration initiates mechanical transmission within the middle ear.

In this air-filled cavity, the ossicular chain (malleus, incus, and stapes) amplifies the sound signal and compensates for

the impedance mismatch between air and the cochlear fluids (Rosowski, 1994). This amplification is essential for ensuring that speech frequencies are transmitted with fidelity, allowing children to access the acoustic cues required to discriminate between similar phonemes. The Eustachian tube maintains pressure equilibrium, ensuring the efficient functioning of the ossicular chain. The vibration transmitted by the stapes is applied to the oval window, initiating the propagation of sound through the cochlear fluids.

The cochlea, spiral-shaped structure located within the temporal bone, contains perilymph and endolymph, fluids whose ionic composition is fundamental to sensory transduction. Vibration at the oval window generates pressure waves that travel along the scala vestibuli and scala tympani, causing displacement of the basilar membrane. This membrane exhibits a tonotopic organisation: higher frequencies are processed at the stiffer basal end, whereas lower frequencies are processed at the more flexible apical region; This mechanism was initially characterized by Békésy (1960) and has since been extensively validated by subsequent research (Robles & Ruggero, 2001). Resting atop the basilar membrane is the organ of Corti, where inner hair cells convert mechanical vibrations into electrical impulses, while outer hair cells modulate sensitivity and frequency selectivity, contributing to the fine discrimination of phonetic contrasts (Dallos, 1992).

Central Auditory Pathways

The action potentials generated by the inner hair cells are transmitted through the cochlear nerve to the cochlear nucleus in the brainstem, where an initial decomposition of the temporal and spectral characteristics of sound takes place (Young & Oertel, 2010). The information then proceeds to the superior olivary complex, which integrates interaural time and intensity differences, enabling the child to locate the source of speech and to orient auditory attention (Grothe et al., 2010). The lateral lemniscus and the inferior colliculus further refine temporal analysis and the detection of acoustic patterns, contributing to the perception of prosody, rhythm, and intonation (Aitkin, 1990).

In the medial geniculate body of the thalamus, the auditory signal is modulated according to attentional demands (Winer, 2006) before reaching the primary auditory cortex in the superior temporal gyrus. In this region, tonotopic organisation is preserved, allowing for detailed analysis of speech frequencies (Hackett, 2011). Secondary and associative auditory areas integrate this information with the language networks of the temporal lobe, supporting word recognition, phonological categorisation, the construction of semantic representations, and the interpretation of syntactic structures (Hickok & Poeppel, 2007). It is precisely this articulation between auditory anatomy and physiology that underpins the processes of language acquisition.

Processes of Language Acquisition

In the first months of life, infants display a remarkable sensitivity to phonetic contrasts, including those that do not belong to their native language. This universal discrimination ability, extensively documented by Werker and Tees (1984), is progressively refined through a process of perceptual specialisation in which the central auditory system adjusts to the phonetic regularities of the language to which the child is exposed (Kuhl, 2004; Kuhl et al., 2008). The formation of stable phonological categories depends on the integrity of the central auditory pathways and on cortical areas responsible for rapid temporal analysis. This is an essential skill for distinguishing phonemes that differ by only a few milliseconds (Tallal, 2004). Lexical acquisition requires the child to associate sound sequences with referents and concepts, a process that relies on the precise perception of the phonological form of words and its integration into memory networks (Hoff, 2013). The quality of phonological representations directly influences vocabulary growth and language comprehension, and is one of the strongest predictors of subsequent lexical development (Bishop, 2014).

Morphosyntactic acquisition involves extracting structural regularities from the sequences of words the child hears. This process is supported by fronto-temporal networks that integrate auditory information with mechanisms of sequential processing, as described by Friederici (2011). Prosody provides essential cues regarding phrase boundaries, informational focus, and syntactic structure, playing a crucial role in speech segmentation and in the comprehension of grammatical relations (Fernald, 1992).

Pragmatic comprehension depends on the integration of linguistic and non-linguistic cues, including intonation, rhythm, and intensity. The auditory perception of suprasegmental elements is essential for interpreting communicative intentions, emotions, and discourse nuances, aspects extensively discussed by Snow (1995).

Within this framework, it becomes evident that the integrity of central auditory processing is a fundamental requirement for language acquisition. When this processing is compromised, as in auditory agnosia (particularly verbal auditory agnosia) the chain linking the acoustic stimulus to linguistic representations is disrupted (Poeppel, 2001). The child may hear sounds but is unable to map them consistently onto phonological units and meanings. The absence or fragility of these early representations reverberates across all levels: limited vocabulary, comprehension difficulties, simplified sentence structures, disruptions in communicative interaction, and significant impact on academic learning (Bishop, 2014). Even non-verbal auditory agnosias, by affecting voice recognition, prosody perception, and the interpretation of social auditory cues, disrupt fundamental aspects of human communication (Belin et al., 2004).

Thus, auditory agnosia constitutes a privileged model for understanding the profound dependence of language on central auditory processing. Analysing this relationship clarifies the neurocognitive mechanisms underlying language acquisition and illuminates the impact that central auditory disturbances can have on child development, communication, and learning.

Auditory Agnosia : Neuropsychological Framework, Subtypes, and Functional Implications

Auditory agnosia is a neuropsychological disorder characterised by the inability to recognise, discriminate, or assign meaning to auditory stimuli, despite preserved peripheral hearing. It is therefore a higher-level central auditory processing deficit resulting from lesions or dysfunctions in cortical and subcortical regions responsible for sound interpretation (Griffiths et al., 2007; Kumar et al., 2012). Unlike peripheral auditory disorders, which impair stimulus detection, auditory agnosia affects comprehension and recognition, highlighting the distinction between hearing and interpreting.

From a neuroanatomical perspective, auditory agnosia is frequently associated with bilateral lesions of the superior temporal lobe, including the primary auditory cortex and, more critically, the secondary and associative auditory areas (Griffths, 2015; Tramo et al., 2002). These regions are responsible for integrating complex acoustic patterns and linking auditory input to linguistic, semantic, and memory networks. Unilateral lesions, particularly in the right hemisphere, may also give rise to specific forms of agnosia, such as phonagnosia, demonstrating that auditory recognition is a distributed and lateralised process (Garrido et al., 2009; Roswandowitz et al., 2014).

The literature distinguishes several subtypes of auditory agnosia according to the functional domain affected.

Verbal auditory agnosia, also referred to as word deafness or pure word deafness, is characterised by an inability to understand spoken language, despite intact hearing and fluent speech production. Individuals detect the presence of speech, but it appears as an undifferentiated sequence of sounds devoid of linguistic value. This subtype is typically associated with bilateral lesions in the perisylvian regions of the temporal lobe, which disrupt the connection between the auditory cortex and the linguistic areas responsible for phonological and semantic decoding (Stewart et al., 2006; Poeppel, 2008).

Non-verbal auditory agnosia refers to difficulty recognising environmental sounds, such as a dog barking, a telephone ringing, or a car engine. In these cases, the deficit is not linguistic but perceptual, affecting the ability to identify sound sources and assign meaning to them. This subtype is frequently linked to right-hemisphere lesions, which play a key role in processing complex acoustic patterns and in the global perception of sound (Clarke & Thiran, 2004; Goll et al., 2010). Musical agnosia, or acquired amusia, corresponds to the inability to recognise melodies, rhythms, or musical structures. Although it may coexist with other auditory deficits, amusia typically results from lesions in the right temporal regions, which are critical for musical processing (Peretz, 2016; Sihvonen et al., 2017).

Finally, phonagnosia is characterised by the inability to recognise familiar voices despite preserved speech comprehension. This deficit highlights the existence of specialised mechanisms for voice recognition that are distinct from linguistic processing (Roswandowitz et al., 2014; Latinus & Belin, 2011).

Regardless of the subtype, auditory agnosia reveals the complexity of central auditory processing and the way in which different dimensions of sound (linguistic, environmental, musical, and identity related) are processed by partially independent neural networks. The distinction between these subtypes demonstrates that auditory recognition is not a unitary process but rather a set of specialised operations that depend on the integrity of specific cortical circuits (Hickok & Poeppel, 2007; Griffiths & Warren, 2004).

The functional implications of auditory agnosia are particularly significant when the disorder occurs during developmental age. In the case of verbal auditory agnosia, the child is exposed to speech but is unable to extract the phonetic regularities necessary to construct stable phonological representations. Without these representations, it becomes impossible to develop a robust lexicon, understand syntactic structures, or acquire morphosyntactic skills (Bishop, 2014; Kuhl, 2010). Language ceases to be an accessible system, not due to a lack of input, but due to an inability to interpret it. This deficit reverberates across all dimensions of linguistic development: reduced vocabulary, comprehension difficulties, persistent grammatical errors, and limitations in spontaneous communication.

Even Non-verbal auditory agnosias can interfere with communicative development, as they compromise prosody perception, voice recognition, and the interpretation of social auditory cues. Prosody is essential for understanding communicative intentions, emotions, and pragmatic nuances (Paulmann & Kotz, 2008); voice recognition contributes to the formation of social bonds and the regulation of interaction (Latinus & Belin, 2011); and the identification of environmental sounds is fundamental for autonomy and safety (Goll et al., 2010).

Thus, auditory agnosia, in any of its forms, can affect not only language but also social interaction, learning, and the child's overall development. It therefore constitutes a privileged model for understanding the relationship between auditory perception and language development. By compromising the ability to assign meaning to auditory stimuli, this disorder highlights the profound dependence of language on central auditory processing. A detailed analysis of auditory agnosia helps clarify the neurocognitive mechanisms that support language acquisition and sheds light on the impact that central auditory disturbances can have on child development, communication, and learning.

Methods

Aim of the Study

This study aims to conduct a systematic literature review regarding the topic of Auditory Agnosia. The primary objective is to evaluate early detection methods for auditory agnosia in children during the language acquisition stage and analyze the impact of this condition on language development, cognition,

and socialization. The secondary objective is to identify gaps in the literature and suggest directions for future research and clinical practices.

Procedure

For the bibliographic research, the databases "PubMed," and "ResearchRabbit" were used. The search terms and the Boolean operators that were used for analysis in the databases are the following: ("Auditory Agnosia" OR "Word Deafness" OR "Pure Word Deafness" OR "NonVerbal Auditory Agnosia") AND ("Language Acquisition" OR "Childhood" OR "Child" OR "Pediatrics" OR "Infant"). The search was conducted in December 2025, gathering all existing articles up to that date.

Inclusion Criteria

Only studies published from 2005 onwards were included. Only articles with full text availability were considered; Studies conducted with children in the language acquisition stage, up to 6 years of age; Studies involving adults were included if they met at least one of the following conditions: 1. presented a longitudinal follow-up originating in childhood; 2. provided retrospective data linking pre-existing auditory deficits to linguistic development; or 3. contributed primary, detailed case reports on the relationship between acquired auditory agnosia and linguistic degradation/reorganization; Only studies that established an explicit relationship between auditory agnosia (regardless of subtype) and linguistic processes were included; Only original studies were selected, encompassing empirical research and detailed case reports that contribute primary data to the field; Articles written in English and/or Portuguese.

Exclusion Criteria

Publications prior to 2005; Studies for which full text access was not available; Research that did not involve children up to 6 years of age was excluded, except in cases where adult participants provided longitudinal or retrospective data relevant to developmental trajectories; Studies that failed to present a formal diagnosis or explicit description of auditory agnosia, or that did not establish a clear and direct relationship between auditory agnosia and linguistic processes; Non-original publications; Articles written in languages other than English or Portuguese.

Results

The identification and selection process, as detailed in the PRISMA Flow Diagram (figure 1), resulted in the final analysis of 25 articles. Initially, records were identified through electronic database searches and screened by title and abstract. After removing duplicates and applying exclusion criteria (such as studies outside the temporal window or lacking a focus on central agnosia), the remaining full texts were assessed for eligibility.

The synthesis of the 25 included studies demonstrates that pediatric auditory agnosia is a complex condition where the timing of intervention is the critical determinant for communicative success. The PRISMA model allowed for the filtering of evidence employing advanced technologies, such as fMRI (Poliva et al., 2015) and longitudinal follow-ups (Kaga et al., 2025), ensuring that the results discussed represent the state-of-the-art in understanding central auditory networks. Table 1 provides a comparative summary of all studies selected through this process.

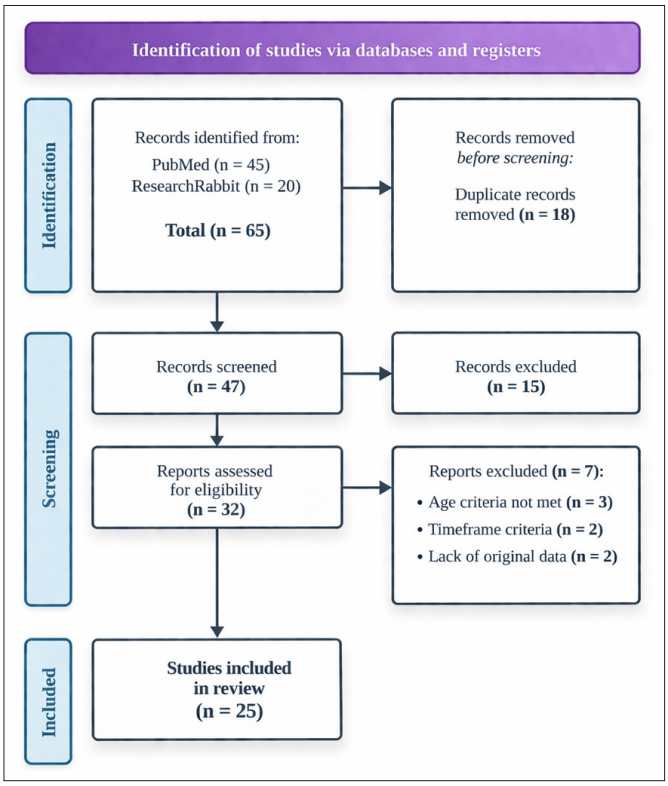

Figure 1: PRISMA 2020 flow diagram for new systematic reviews which included searches of databases and registers only (Page et al., 2021).

Methodological Quality Assessment

The methodological quality of the 25 studies selected via the PRISMA protocol was rigorously evaluated using the JBI (Joanna Briggs Institute) checklists for case reports/series and the CASP tools for observational studies.

The overall quality of the evidence was rated as moderate to high. Adherence to the PRISMA guidelines minimized selection bias and ensured that the clinical data possesses sufficient scientific validity to support the clinical conclusions of this review. The consistency found in neurophysiological biomarkers (such as the absence of MLR in the presence of normal ABR) across different study designs reinforces the reliability of the integrated findings. This methodological rigor underscores that the identified auditory-linguistic deficits are rooted in specific central network disruptions rather than peripheral hearing loss.

Discussion

The understanding of auditory-linguistic disorders has undergone a fundamental paradigm shift, moving beyond an exclusive focus on the integrity of the peripheral auditory system to investigate the complexity of cortical and subcortical neural networks. The body of evidence reviewed in this work

corroborates this transition, illuminating the multifactorial and often heterogeneous nature of the mechanisms underlying sound perception and language processing. Although the included studies employ diverse methodologies, ranging from case reports with functional fMRI mapping to genetic and longitudinal analyses, the synthesis of these data reveals converging themes that challenge conventional clinical assessments and point toward the need for a dynamic view of the neurobiology of hearing.

In this section, the literature findings are integrated and discussed across seven thematic domains, contextualizing the reviewed studies within contemporary theoretical frameworks.

Central Auditory Processing Alterations and Auditory Agnosias

The body of evidence reviewed converges on the fundamental principle that central auditory dysfunction can manifest devastatingly even when peripheral audiometry remains within normal limits. This observation reinforces the clinical axiom that cochlear integrity and "bottom-up" signal input do not, by themselves, guarantee cortical perception or meaningful sound processing (Pillion, 2012; Hattiangadi et al., 2005). A granular analysis of the literature reveals that the specific phenotype of the deficit is strictly tied to the anatomical level of the lesion within the auditory hierarchy. At the subcortical level, disruption in the inferior colliculus, which serves as a mandatory midbrain relay for nearly all ascending auditory fibers, severely compromises the ability to resolve temporal features of sound. As demonstrated by Poliva et al. (2015), damage to this structure prevents bilateral integration and fine frequency discrimination, resulting in a degraded signal reaching the cortex that manifests as generalized auditory agnosia, even though the ABR remains normal as it reflects activity at levels below the colliculus.

Table 1: Synthesis of clinical and neurophysiological evidence from the included studies (n=25) on pediatric auditory agnosia.

Author(s)	Year	Study Type	Sample	Objective	Main Findings	Conclusions
Hattiangadi et al.	2005	Case Report	1 child	Characterize auditory agnosia after TBI	Peripheral hearing preserved (OAE, ABR), absent MLR, severe speech deficits	Cortical auditory agnosia can follow TBI; central tests are essential
Metz Lutz	2009	Review	--	Review auditory function in CSWS/LKS	Verbal agnosia is central; phonological deficit affects verbal memory	LKS involves superior temporal dysfunction; phonological deficits explain deterioration
Chou et al.	2010	Case Report	1 child	Medulloblastoma presenting with PWD	Near normal peripheral hearing, severe verbal deficit	Tumors can cause PWD via central auditory pathways
Zhang et al.	2011	Case Report	1 adult	Auditory agnosia after chronic hydrocephalus	Auditory cortices intact; auditory radiations destroyed; music preserved	Hydrocephalus can cause auditory agnosia; secondary pathways may sustain music
Pillion	2012	Case Series	3 patients	Describe speech processing deficits from neural lesions	Case 1: ANSD; Case 2: inferior colliculus lesions; Case 3: cortical agnosia	Normal audiogram does not guarantee normal processing; ABR/MLR essential
Van Bogaert et al.	2012	Case Series	3 children	LKS with initially normal EEG	EEG normal at onset; IEDs appear months later	Normal EEG does not exclude LKS
Devinsky et al.	2014	Case Series	2 children	LKS with episodic auditory agnosia	Rapid response to diazepam + steroids	Combination therapy may be more effective than monotherapy

Baird et al.	2014	Case Report	1 young musician	Apperceptive music agnosia after surgery	Selective preservation of beat; melodic deficit	Rhythm–meter dissociation; anterior STG involved in metric processing
Furushima et al.	2015	Case Series	3 Children	Characterize early auditory loss in ALD	Normal audiometry & ABR; severe discrimination deficits	ALD can cause early central auditory agnosia
Poliva et al.	2015	Case Report (fMRI)	1 adult	Map auditory cortical fields in agnosia from IC trauma	Sounds activated caudal hA1 and posterior STG, but not rostral hR or anterior STG	Agnosia reflects ventral stream dysfunction; streams are segregated at the primary cortex
Kaga et al.	2015	Case Report	1 adult	Study pathophysiology of cortical deafness and sensory loss	Total hearing loss with normal OAE/ ABR; bilateral lesions in auditory & postcentral gyri	Cortical deafness can coexist with somatosensory loss; peripheral tests don't rule out cortical injury
Lopez Soto et al.	2016	Case Report	2 children	Diagnose cortical hearing loss via LLEP	Normal ABR, immature LLEP	LLEP useful for diagnosing cortical immaturity
Szmuda et al.	2016	Systematic Review	101 studies	Evaluate DTI usefulness in pediatric epilepsy	DTI useful in TLE and surgical planning	Should be included in selected cases
Riccio et al.	2016	Case Series	14 children	Characterize neurocognitive profiles in LKS	57% auditory deficits; 50% verbal memory deficits	LKS has broad cognitive impact; deficits persist
Necula et al.	2017	Case Report	1 young adult	Verbal agnosia after herpes encephalitis	Bilateral temporal lesions; normal peripheral hearing	Early encephalitis can cause severe verbal agnosia
Maffei et al.	2017	Case Report	1 woman	PWD after left temporal stroke	Environmental sounds & music preserved; verbal deficit only	PWD can occur with unilateral lesions; functional disconnection involved
Strehlow et al.	2019	Genetic Study	248 individuals	Genotype–phenotype correlations in GRIN2A	MisTMD = GOF & severe phenotype; misATD/LBD = LOF & mild phenotype	Functional model predicts severity and guides therapy
Mir et al.	2020	Case Report	1 child	Test memantine as targeted therapy in GRIN2A	Seizure free after memantine	Example of precision medicine in genetic epilepsy
Klarendić et al.	2021	Case Report	1 woman	Auditory agnosia with anosognosia after strokes	General agnosia: verbal, environmental, musical; staged recovery	Written language preserved; important for differential diagnosis

Gwilliams et al.	2022	MEG Study	21 adults	Investigate neural coding of phonemes	Brain encodes 3 phonemes in parallel; position-invariant coding	Explains how brain maintains phoneme order despite overlapping activity
Porcar Gozalbo et al.	2024	Cross-sectional	140 children	Impact of hearing loss type on development	Bilateral/severe losses worst; early diagnosis predicts outcomes	Early auditory & language intervention is essential
Okahara et al.	2024	Retrospective	21 TLE patients	Evaluate passive fMRI for language mapping	95.2% concordance with Wada test; works in ID patients	Passive fMRI is reliable for presurgical language mapping
Ngoh et al.	2025	Cohort Study	52 LKS patients	Clinical & genetic characterization of LKS	Early onset predicts worse outcomes; GRIN2A in 15.5%	Early intervention crucial; EEG not reliable alone; new genes identified
Doucet et al.	2025	Resting-state fMRI	22 DLD children	Identify whole-brain FC abnormalities in DLD	Widespread reduced FC in SM, CC, DM networks	DLD involves broad neural dysfunction beyond language areas
Kaga et al.	2025	Long-term follow up	1 patient	Follow progression from agnosia to cortical deafness	Thresholds progressed to profound loss; ABR normal at age 29; bilateral cortical damage	Early agnosia can evolve into permanent cortical deafness via retrograde degeneration

Abbreviations

ABR : Auditory Brainstem Response;
DPOAE: Distortion Product Otoacoustic Emissions;
fMRI : Functional Magnetic Resonance Imaging;
MLR : Middle-Latency Response;
LLEP : Long-Latency Evoked Potentials;
DTI : Diffusion Tensor Imaging;
EEG : Electroencephalogram;
LKS : Landau-Kleffner Syndrome

Moving up the neural hierarchy, lesions in the auditory radiations represent a "disconnection syndrome" where the primary auditory cortex may remain physically intact, but the absence of functional thalamic afferents, as seen in cases of chronic hydrocephalus (Zhang et al., 2011) or severe encephalitis (Kaga et al., 2025), leads to cortical deafness. This scenario highlights that perception requires not only the presence of cortical tissue but a preserved physical and functional link to the thalamus. In cases of extensive bilateral destruction of Heschl's gyri, cortical deafness may be total. It is crucial to note that such vascular or inflammatory insults rarely occur in isolation; due to neuroanatomical proximity to the postcentral gyrus and vestibular processing areas, the condition can evolve into a multimodal sensory loss (Kaga et al., 2015). In these situations, the patient presents not only deaf to sound but also with a loss of somatosensory and vestibular sensations, illustrating the compromise of a broader sensory "hub." The central paradox of these alterations lies in the preservation of peripheral biomarkers: the presence of normal distortion product otoacoustic emissions (DPOAE) confirms the health of the outer hair cells, while a normal ABR validates the integrity of the auditory nerve and lower brainstem. It is precisely this diagnostic gap between the functional periphery and the inability to hear that defines the domain of agnosias and central auditory processing disorders.

Language, Phonology, and the Segregation of Auditory Streams

The literature on Landau-Kleffner Syndrome (LKS) and acquired aphasias has traditionally highlighted the high vulnerability of the superior temporal cortex and its associated phonological networks. However, contemporary functional neuroimaging has refined this view by demonstrating that auditory processing is not a monolithic event but is organized into segregated functional streams as early as the primary auditory cortex. This dual-stream model consists of a ventral stream (responsible for the recognition of auditory objects or the "what" of a sound) and a dorsal stream, which manages spatial localization and the "where" of a stimulus.

According to Poliva et al. (2015), auditory agnosia and pure word deafness appear to specifically reflect a localized dysfunction of the ventral stream, particularly involving the rostral sub-area of the primary auditory cortex (hR) and the anterior superior temporal gyrus (STG). In such cases, the dorsal stream may remain remarkably functional. This explains the clinical phenomenon in which a patient can accurately localize a sound source in space or detect its movement, despite being completely unable to recognize the sound's identity or comprehend spoken words (Poliva et al., 2015).

Furthermore, the modular nature of these networks is reinforced by observed dissociations between different types of meaningful sounds. For instance, the preservation of musical abilities in the face of severe speech perception deficits (Maffei et al., 2017) suggests that, within higher-order auditory pathways, neural resources are partitioned. This modularity implies that "auditory-linguistic" disorders are often the result of specific network disconnections rather than a generalized loss of auditory intelligence, where the brain's ability to map sound to meaning (ventral) is severed, while its ability to map sound to action or space (dorsal) remains intact.

Genetic and Neurobiological Contributions

Recent genetic research has revolutionized the understanding of auditory-linguistic disorders by identifying the central role of the GRIN2A gene, which encodes the GluN2A subunit of NMDA receptors (Strehlow et al., 2019). These receptors are fundamental for synaptic plasticity and the development of language networks. The literature now establishes a clear genotype-phenotype correlation: gain-of-function variants, which increase receptor activity, tend to manifest in severe phenotypes such as Landau-Kleffner Syndrome and epileptic encephalopathy with continuous spike-and-wave during sleep (CSWS), resulting in profound language regressions (Ngoh et al., 2025). In contrast, loss-of-function variants are generally associated with milder forms of developmental language disorder and benign childhood epilepsies (Strehlow et al., 2019).

This neurobiological insight has enabled a transition toward precision medicine, where pharmacotherapy is guided by the underlying molecular mechanism. A paradigmatic example is the use of memantine, a non-competitive NMDA receptor antagonist. In cases where the GRIN2A mutation causes pathological overactivation of receptors, memantine acts by blocking the excess excitatory signaling, resulting not only in better seizure control but also in partial recovery or stabilization of the neurocognitive and linguistic profile (Mir et al., 2020). This approach demonstrates that language regression in these syndromes is not merely an electrical (epileptic) phenomenon but a treatable synaptic dysfunction, where restoring neurochemical balance can mitigate the impact on central auditory networks.

Functional Connectivity and Neural Language Networks

Contemporary functional neuroimaging has significantly expanded the scope of auditory-linguistic disorders, enabling a transition from the analysis of isolated areas to the assessment of distributed neural networks. A crucial methodological advancement lies in the use of passive fMRI protocols, which have proven to be effective and less demanding tools for mapping language lateralization, especially in populations with severe cognitive deficits or cooperation difficulties, where behavioral hearing tests and the Wada test are often unfeasible (Okahara et al., 2024). These studies demonstrate that even in the absence of active tasks, the functional organization of language networks can be identified through connectivity biomarkers, offering a safe window for pre-surgical and diagnostic evaluation.

Furthermore, the understanding of conditions such as Developmental Language Disorder (DLD) has been profoundly altered by the analysis of resting-state connectivity. Currently, DLD is understood not as an isolated linguistic deficit, but as a distributed network disorder involving systemic dysfunction. As evidenced by Doucet et al. (2025), children with DLD exhibit reduced functional connectivity that extends far beyond the classical Broca's and Wernicke's areas, affecting sensorimotor, cognitive control, and default mode networks.

This altered neural architecture suggests that difficulties in language acquisition may reflect the brain's inability to integrate information across global motor and cognitive domains. Thus, the pathology is no longer viewed as a "lesion" in a specific location but is interpreted as a failure in the synchrony and integrity of cortical networks, where inefficient communication between multiple neural systems compromises the fluid development of auditory-linguistic skills (Doucet et al., 2025; Okahara et al., 2024).

Language Development and the Impact of Hearing Loss

Early detection and timely intervention remain the most robust predictors of successful linguistic development in children with hearing deficits (Porcar-Gozalbo et al., 2024). However, recent literature has introduced a critical perspective on the evolutionary nature of these disorders. A fundamental finding from long-term longitudinal studies reveals that auditory agnosia in childhood may not be a static condition, but rather a clinical framework that can progress to permanent cortical deafness in adulthood (Kaga et al., 2025). This late-onset deterioration is attributed to the retrograde degeneration of the auditory radiations and thalamocortical pathways, where a lack of functional processing or chronic tissue damage results in the progressive atrophy of the nerve fibers connecting the thalamus to the cortex.

This progression phenomenon, observed in follow-ups spanning more than three decades, underscores that while brain plasticity is particularly potent in childhood and allows for initial compensations, it can be limited or even reversed by long-term secondary degenerative processes (Kaga et al., 2025). Consequently, the stability of auditory thresholds in childhood does not guarantee the maintenance of function in adulthood, especially in cases of sequelae from encephalitis or severe vascular lesions. This evidence reinforces the need for continuous audiological and neurophysiological monitoring, suggesting that intervention strategies must be dynamic and

adaptive to mitigate the impact of the progressive loss of central connectivity within the auditory system (Kaga et al., 2025; Porcar-Gozalbo et al., 2024).

Neurophysiological Assessment and Auditory Biomarkers

The scientific literature highlights the critical utility of short, middle, and late latency auditory evoked potentials (ABR, MLR, and LLEP, respectively) in identifying central deficits that elude conventional audiometry (Lopez-Soto et al., 2016). While the Auditory Brainstem Response (ABR) is essential for validating the integrity of the pathway up to the midbrain, the presence of a normal ABR in patients with functional hearing inability points to a pathology located above this level. In this context, the Middle Latency Response (MLR) emerges as a robust biomarker of thalamocortical dysfunction; its absence or severe attenuation is a reliable indicator that the auditory signal is not being effectively transmitted from the thalamus to the primary auditory cortex, regardless of peripheral hearing status (Hattiangadi et al., 2005).

In addition to electrophysiological tests, functional mapping via fMRI has emerged as a precision diagnostic tool to differentiate selective agnosias. By identifying "non-reactive" cortical areas to specific sound stimuli, such as the rostral sub-area (hR) of the auditory cortex, clinicians can obtain a functional biomarker of processing disruption in the ventral stream (Poliva et al., 2015). Complementarily, Diffusion Tensor Imaging (DTI) allows for the visualization of the structural integrity of the auditory radiations and white matter connectivity, providing physical evidence of demyelination or axonal degeneration (Szmuda et al., 2016). The integration of these biomarkers—functional (fMRI), structural (DTI), and neurophysiological (MLR/LLEP)—is what currently allows for overcoming the limitations of diagnostics based solely on behavioral hearing, offering an objective characterization of cortical deafness and agnosias (Lopez-Soto et al., 2016; Poliva et al., 2015).

Clinical and Therapeutic Implications

The integrated evidence from this review points toward three fundamental clinical implications that should guide the management of complex auditory-linguistic disorders. First, there is an imperative need for prolonged clinical and neurophysiological follow-up, as the absence of pathological findings in initial exams can be misleading. As demonstrated by Van Bogaert et al. (2012), a normal EEG in children with language regression does not exclude a diagnosis of Landau-Kleffner Syndrome (LKS), given that characteristic epileptiform discharges may manifest only months after the onset of linguistic symptoms. Therefore, clinical vigilance must be continuous, preferably utilizing long-term sleep EEG to capture spike-and-wave activations that would go unnoticed in routine examinations.

Secondly, regarding pharmacological interventions, the literature suggests the superiority of combined approaches over monotherapy in cases of epileptic encephalopathies with language impact. The synergistic use of benzodiazepines (such as high-dose diazepam at sleep onset) and corticosteroids has shown more effective results in the rapid remission of discharges and the stabilization of auditory comprehension compared to the isolated use of anticonvulsants (Devinsky et al., 2014). This multimodal strategy aims not only to control epilepsy but also to protect the plasticity of auditory networks during critical developmental windows.

Finally, the findings underscore the necessity for adaptive interventions throughout life. The discovery that the auditory profile may deteriorate decades after the initial insult due to late neural degeneration processes (Kaga et al., 2025) necessitates a shift in patient management: successful rehabilitation in childhood should not lead to permanent discharge. Instead, the therapeutic plan must provide for periodic reassessments in adulthood to monitor the integrity of the auditory radiations and adjust support strategies (whether through technical aids, psychosocial support, or cognitive rehabilitation), ensuring that quality of life and communicative functionality are preserved in the face of the neurodegenerative evolution of the central auditory system (Kaga et al., 2025; Van Bogaert et al., 2012).

Study Limitations

Although the reviewed literature provides valuable insights into the neurobiological and clinical dimensions of auditory-linguistic disorders, several limitations must be acknowledged to contextualize the strength and generalizability of the findings. These constraints arise from methodological variability, sample characteristics, and the vast etiological diversity of the conditions examined.

Methodological Heterogeneity and Comparative Challenges: One of the primary limitations lies in the disparity of study designs among the analyzed articles. The coexistence of single-case reports, genetic cohort studies, and systematic reviews makes direct data comparison difficult. For instance, while fMRI studies (Poliva et al., 2015) offer granular detail regarding cortical activation, they do not allow for the same population-based prevalence as cross-sectional studies (Porcar-Gozalbo et al., 2024). This heterogeneity restricts the ability to draw generalizable inferences and limits the establishment of robust causal relationships between central auditory deficits and global neurocognitive functioning.

Sample Representativeness and Control Groups: Many findings rely on isolated clinical cases or very small samples, which compromises external validity. Although case studies are fundamental for describing rare phenomena, such as the progression from agnosia to cortical deafness over decades (Kaga et al., 2025), they do not allow for conclusions to be extrapolated to the general population. Furthermore, the frequent absence of matched control groups in neurophysiological studies limits the interpretation of results, making it difficult to distinguish between individual variations and specific pathological markers.

Fragmentation of Assessment Instruments: The lack of standardization in auditory and linguistic assessment batteries constitutes a significant obstacle. Studies utilize a diverse range of instruments, from evoked potentials (ABR, MLR, LLEP) to language scales such as the CELF-5 or temporal processing tests. This methodological diversity complicates

the meta-analysis of data and may introduce biases related to the sensitivity and specificity of each instrument. Without standardized protocols, the consistency of auditory biomarkers across different research centers remains limited.

Etiological Variability and Unified Models: Finally, the high etiological diversity of the conditions examined, which ranges from GRIN2A mutations and herpetic encephalitis sequelae to hydrocephalus and tumors, makes it difficult to identify common pathophysiological mechanisms. Each etiology possesses its own temporal dynamics and structural impact (Kaga et al., 2015). This dispersion complicates the construction of unified theoretical models for auditory agnosia, as the disruption of neural networks can occur through biological pathways as distinct as retrograde neurodegeneration or metabolic demyelination.

Conclusion

The synthesis of the analyzed literature highlights that the timing of identification and intervention is the most critical determinant in the evolution of auditory-linguistic disorders. The findings demonstrate that these pathologies are not static entities but rather the result of a dynamic and complex interaction between central processing deficits, linguistic impairments, and neurobiological factors. This perspective confirms that functional hearing transcends cochlear integrity, requiring an analysis of the cortical and subcortical networks that sustain higher-order processing (Metz-Lutz, 2009; Doucet et al., 2025).

The gathered evidence reinforces that severe central alterations can coexist with normal audiometric thresholds. The functional distinction between the ventral (recognition) and dorsal (localization) streams reveals that auditory agnosia can be selective, preserving spatial perception while obliterating the meaning of sound (Poliva et al., 2015). Furthermore, long-term monitoring introduces a crucial clinical warning: the possibility of retrograde degeneration of the auditory radiations, which can convert childhood agnosia into permanent cortical deafness in adulthood (Kaga et al., 2025). This finding underscores the need to integrate neurophysiological biomarkers (such as MLR and LLEP) and functional neuroimaging to monitor neural connectivity over decades.

In parallel, advances in genetics, particularly in the study of the GRIN2A gene, pave the way for precision medicine where pharmacological treatment can be targeted at the underlying molecular mechanism, mitigating linguistic regressions that were previously considered untreatable (Strehlow et al., 2019; Mir et al., 2020). The etiological diversity observed, ranging from herpetic encephalitis to genetic mutations and vascular lesions, calls for integrated theoretical models and multimodal assessment protocols that link neurology, genetics, and audiology.

In summary, the conclusions of this review underscore the urgent need for more sensitive diagnostic tools and dynamic interventions. Progress in this field will depend on the ability of clinical practice to respond to the uniqueness of each neurobiological profile, ensuring that brain plasticity is maximized and that secondary degenerative processes are identified and mitigated early.

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