Neurocase (1999) Vol. 5, pp. 379–406

PREVIOUS CASES

The Auditory Agnosias

Jon S. Simons and **Matthew A. Lambon Ralph**
MRC Cognition and Brain Sciences Unit, Cambridge

Auditory agnosia refers to the defective recognition of auditory stimuli in the context of preserved hearing. There has been considerable interest in this topic for over a hundred years despite the apparent rarity of the disorder and potential diagnostic confusion with deafness or even Alzheimer's disease (Mendez and Rosenberg, 1991—see Case P593 below). Following Lissauer's (1890) distinction between 'apperceptive' and 'associative' forms of visual object agnosia, disorders of sound recognition have been divided between impaired perception of the acoustic structure of a stimulus, and inability to associate a successfully perceived auditory representation with its semantic meaning (Vignolo, 1982).

Much research has centred on the 'apperceptive' form of auditory agnosia, although the study of such disorders has not been aided by terminological differences in the literature. Some authors have used the term 'auditory agnosia' to refer to a generalized disorder affecting the perception of all types of auditory stimuli, from non-verbal sounds to speech and music (e.g., Miceli, 1982). Others have employed separate names to highlight specific deficits, using 'auditory agnosia' to refer to impairment of non-verbal, environmental sounds (Albert *et al.*, 1972—see Case P565 below), 'amusia' to escribe deficits in the processing of melodies (e.g., Peretz *et al.*, 1994—see Case P601 below), and 'pure' word deafness for disorders of speech recognition (e.g., Albert and Bear, 1974; Saffran *et al.*, 1976; Coslett *et al.*, 1984—see Cases P605 and P569 below). While 'pure' word deafness is perhaps the most commonly used label, it has been noted for some time that the disorder is rarely pure—in the majority of cases there is some concomitant impairment of non-verbal sound processing (Hemphill and Stengel, 1940—see Case P583 below; Nielsen, 1946; Wohlfart *et al.*, 1952—see Case P613 below). Furthermore, many cases are not even free of aphasia (as implied by the original use of the term 'pure': Auerbach *et al.*, 1982—see Case P567 below; Vignolo, 1982).

Analysis of impaired speech perception in word deafness suggests that certain speech sounds may be affected more than others (Saffran *et al.*, 1976—see Case P605 below). Auditory comprehension can be improved by lip-reading (Shindo *et al.*, 1991—see Case P606 below) and context (Saffran *et al.*, 1976). While impaired speech recognition normally occurs in the presence of generalized auditory agnosia, some patients have been described with an apparently language-specific disorder (Auerbach *et al.*, 1982). Franklin (1989—see Case P574 below) highlighted five different levels of language-specific impairment that might give rise to poor spoken comprehension. One of these, word meaning deafness, is a form of 'associative' auditory agnosia that has fascinated researchers ever since Bramwell first described the disorder at the end of the nineteenth century (see Ellis, 1984).

To be a classic case of word meaning deafness, a patient should have preserved repetition, phoneme discrimination and lexical decision, but impaired comprehension from spoken input alone (comprehension is normal for written words and pictures: Franklin *et al.*, 1996; Kohn and Friedman, 1986—see Cases P576 and P589 below). Spoken comprehension is graded by imageability/concreteness—words referring to concrete exemplars are more likely to be understood than abstract concepts—and errors are generally semantically related (Franklin *et al.*, 1994, 1996—see Cases P575 and P576 below). Word meaning deafness would seem, therefore, to be a language-specific deficit of the highest order although Tyler and Moss (1997—see Case P611 below) have suggested that, given the speed and accuracy required in speech perception, this behavioural pattern may be the natural by-product of a more generalized (earlier) auditory processing impairment.

Auditory agnosias are often associated with bilateral, or unilateral left, lesions of the superior temporal region (Auerbach *et al.*, 1982; Varney and Damasio, 1986) although occasional cases have been described with unilateral right temporal involvement (Roberts *et al.*, 1987—see Case P603 below). By far the most common cause is vascular accident but some cases have been reported following encephalitis (Arias *et al.*, 1995—see Case P566 below), head injury (Franklin, 1989—see Case P574 below) and slow progressive atrophy (Otsuki *et al.*, 1998—see Case P598 below). Childhood auditory agnosia has been described in association with epileptogenic, temporal lobe abnormalities on EEG (Kale *et al.*, 1995; Stein and Curry, 1968—see Cases P586 and P608) that can improve with anticonvulsant medication (Pearce and Darwish, 1984—see Case P600 below). A comprehensive review of the current state of the art in this area is provided by Griffiths *et al.* (1999) in this issue of *Neurocase*.

References

Albert ML, Bear D. Time to understand: a case study of word deafness with reference to the role of time in auditory comprehension. Brain 1974; 97: 373–84.

Albert ML, Sparks R, Von Stockert T, Sax D. A case study of auditory agnosia: linguistic and non-linguistic processing. Cortex 1972; 8: 427–43.

Arias M, Requena I, Ventura M, Pereiro I, Castro A, Alverez A. A case of deaf-mutism as an expression of pure word deafness: neuroimaging and electrophysiological data. European Journal of Neurology 1995; 2: 583–5.

Auerbach SH, Allard T, Naeser M, Alexander MP, Albert ML. Pure word deafness. Analysis of a case with bilateral lesions and a defect at the prephonemic level. Brain 1982; 105: 271–300.

Coslett HB, Brashear HR, Heilman KM. Pure word deafness after bilateral primary auditory cortex infarcts. Neurology 1984; 34: 347–52.

Ellis AW. Introduction to Bramwell's (1897) case of word meaning deafness. Cognitive Neuropsychology 1984; 1: 245–58.

Franklin S. Dissociations in auditory word comprehension; evidence from 9 fluent aphasic patients. Aphasiology 1989; 3: 189–207.

Franklin S, Howard D, Patterson K. Abstract word meaning deafness. Cognitive Neuropsychology 1994; 11: 1–34.

Franklin S, Turner J, Lambon Ralph MA, Morris J, Bailey PJ. A distinctive case of word meaning deafness? Cognitive Neuropsychology 1996; 13: 1139–62.

Griffiths TD, Rees A, Green GR. Disorders of human complex sound processing. Neurocase 1999; 5: 365–78.

Hemphill RE, Stengel E. A study of pure word-deafness. Journal Neurology and Psychiatry 1940; 3: 251–62.

Kale U, El-Naggar M, Hawthorne M. Verbal auditory agnosia with focal EEG abnormality: an unusual case of a child presenting to an ENT surgeon with 'deafness'. The Journal of Laryngology and Otology 1995; 109: 431–2.

Kohn SE, Friedman RB. Word-meaning deafness: a phonological-semantic dissociation. Cognitive Neuropsychology 1986; 3: 291–308.

Lissauer H. Ein Fall von Seelenblindheit nebst einem Beitrage zur Theorie derselben. Archiv für Psychiatrie und Nervenkrankheiten 1890; 21: 222–70.

Mendez MF, Rosenberg S. Word deafness mistaken for Alzheimer's disease: differential characteristics. Journal of the American Geriatrics Society 1991; 39: 209–11.

Miceli G. The processing of speech sounds in a patient with cortical auditory disorder. Neuropsychologia 1982; 20: 5–20.

Nielsen JM. Agnosia, apraxia, aphasia. Their value in cerebral localization. New York: Hoeber, 1946.

Otsuki M, Soma Y, Sato M, Homma A, Tsuji S. Slowly progressive pure word deafness. European Neurology 1998; 39: 135–40.

Pearce PS, Darwish H. Correlation between EEG and auditory perceptual measures in auditory agnosia. Brain and Language 1984; 22: 41–8.

Peretz I, Kolinsky R, Tramo M, Labrecque R, Hublet C, Demeurisse G *et al.* Functional dissociations following bilateral lesions of auditory cortex. Brain 1994; 117: 1283–1301.

Roberts M, Sandercock P, Ghadiali E. Pure word deafness and unilateral right temporo-parietal lesion: a case report. Journal of Neurology, Neurosurgery and Psychiatry 1987; 50: 1708–9.

Saffran EMM, Marin OS, Yeni-Komshian GH. An analysis of speech perception in word deafness. Brain and Language 1976; 3: 209–28.

Shindo M, Kaga K, Tanaka Y. Speech discrimination and lip reading in patients with word deafness or auditory agnosia. Brain and Language 1991; 40: 153–61.

Stein LK, Curry FK. Childhood auditory agnosia. Journal of Speech and Hearing Disorders 1968; 33: 361–70.

Tyler LK, Moss HE. Imageability and category-specificity. Cognitive Neuropsychology 1997; 14: 293–318.

Varney N, Damasio H. CT scan correlates of sound recognition defect in aphasia. Cortex 1986; 22: 483–6.

Vignolo LA. Auditory agnosia. Philosophical Transactions of the Royal Society of London B: Biological Sciences 1982; 298: 49–57.

Wohlfart G, Lindgren A, Jernelius B. Clinical picture and morbid anatomy in a case of 'pure word deafness'. Journal of Nervous and Mental Disease 1952; 116: 818–27.

A case study of auditory agnosia: linguistic and non-linguistic processing

M. L. Albert, R. Sparks, T. von Stockert and **D. Sax**

Abstract

The authors have evaluated at length an individual who had as a major neurological defect an impaired ability to recognize the nature of heard non-verbal sounds, that is, auditory agnosia. This case provides additional clinical support for the theoretical position that there may be two central auditory processing mechanisms, one treating linguistic inputs and the other non-linguistic inputs.

Journal

Cortex 1972; 8: 427–43

Neurocase Reference Number:

P565

Primary diagnosis of interest

Auditory agnosia, auditory neglect, aphasia

Author's designation of case

Not mentioned

Key theoretical issue

- The auditory agnosia seen in this patient supports the view that there are separate verbal and non-verbal auditory processing systems

Key words: auditory agnosia

Scan, EEG and related measures

EEG, radioisotope scan

Standardized assessment

Seashore Measures of Musical Talents, Boston Diagnostic Aphasia Examination, Wechsler Adult Intelligence Scale

Other assessment

Audiometric screening, spontaneous speech, naming, comprehension of spoken language, Repetition tests, reading, writing, singing, calculation, sound localization, dichotic sounds, recognition of non-verbal sounds

Lesion location

- Bilateral posterior cortex

Lesion type

Not known

Language

English

A case of deaf-mutism as an expression of pure word deafness: neuroimaging and electrophysiological data

M. Arias, I. Requena, M. Ventura, I. Pereiro, A. Castro and **A. Alverez**

Abstract

The authors report a case of pure word deafness, clinically expressed as deaf-mutism in a 17-year-old girl, who was affected with encephalitis when she was 18 months old and had not acquired language skills. Actually, physical examination revealed buccolingual apraxia and absence of spontaneous speech, auditory comprehension, repetition and denomination, whereas perception of non-verbal sounds was preserved. The seven waves of brainstem auditory evoked responses (BAER) were present with normal latencies; middle latency responses (MLR) were also normal. Magnetic resonance imaging (MRI) showed bilateral temporoparietal lesions. This case proves that lesions which may give rise to word deafness, when they occur in prelingual age, can determine a peculiar deaf-mutism clinical picture.

Journal

European Journal of Neurology 1995; 2: 583–5

Neurocase Reference Number:

P566

Primary diagnosis of interest

Prelingual, bilateral temporoparietal lesions

Author's designation of case

Not mentioned

Key theoretical issue

- The impact of prelingual, bilateral temporoparietal lesions on the subsequent development of spoken language comprehension and auditory agnosia

Key words: pure word deafness; deaf-mutism; childhood encephalitis

Scan, EEG and related measures

EEG, MRI, Brainstem Auditory Evoked Responses (BAER)

Standardized assessment

Pure-tone audiogram

Other assessment

Clinical assessment of non-verbal sound recognition, and buccolingual apraxia

Lesion location

- Bilateral corticosubcortical temporoparietal lesions (MRI)

Lesion type

Encephalitis

Language

English

Pure word deafness. Analysis of a case with bilateral lesions and a defect at the prephonemic level

S. H. Auerbach, T. Allard, M. Naeser, M. P. Alexander and **M. L. Albert**

Abstract

The authors have demonstrated that the deficit in speech perception in a patient with pure word deafness is secondary to a prephonemic temporal auditory acuity disorder. They delineated the nature of the auditory processing deficit in the patient with bilateral lesions and then demonstrated the presence of a predicted deficit in phonemic discrimination. This pattern is comparable to previous cases with bilateral lesions and distinct from other cases with unilateral lesions. Review of previous reports suggests that there are two distinct types of pure word deafness: type 1, in which the deficit is prephonemic and related to a temporal auditory acuity disorder, and type 2, a form that is independent of a temporal auditory acuity disorder, and has a deficit in linguistic discrimination that does not adhere to a prephonemic pattern [Denes and Semenza, 1975 (P571); Saffran *et al.*, 1976 (P605)]. The former has been associated with bilateral temporal lobe lesions [Naeser, 1974 (presented at the 12th Annual Academy of Aphasia Meetings, Warrenton, VA, October 1974); Chocholle *et al.*, Neuropsychologia 1975; 13: 162–72], the latter with left unilateral lesions [Denes and Semenza, 1975 (P571); Saffran *et al.*, 1976 (P605)]. The first form is an apperceptive disorder, whereas the second represents a higher disorder in phonemic discrimination and may be considered a fragment of Wernicke's aphasia.

Journal

Brain 1982; 105: 271–300

Neurocase Reference Number:

P567

Primary diagnosis of interest

Pure word deafness

Author's designation of case

Not mentioned

Key theoretical issue

- Differentiation of pure word deafness from general auditory agnosia

Key words: pure word deafness; auditory agnosia

Scan, EEG and related measures

CT, isotope brain scan, EEG

Standardized assessment

Spreen and Benton Sound Recognition Test, Brainstem Auditory Evoked Responses

Other assessment

Pure tone threshold audiometry, dichotic listening studies, cortical auditory evoked potentials, temporal auditory acuity measures, natural and synthetic speech sound tests, music tests, sound discrimination, recognition of environmental sounds

Lesion location

- Right middle cerebral infarct, extending from the right middle and posterior temporal region to the parietal lobe, including the postcentral and supramarginal gyri; and a small, older non-enhancing left posterior temporal lobe infarct including part of Wernicke's area.

Lesion type

CVA

Language

English

Chronic auditory agnosia following Landau–Kleffner syndrome: a 23 year outcome study

K. Baynes, J.A. Kegl, D. Brentari, C. Kussmaul and **H. Poizner**

Abstract

The authors report on a 27-year-old woman with chronic auditory agnosia following Landau–Kleffner Syndrome diagnosed at age $4\frac{1}{2}$ years. She grew up in the hearing/speaking community with some exposure to manually coded English and American Sign Language. Manually coded (signed) English is her preferred mode of communication. Comprehension and production of spoken language remain severely compromised. Disruptions in auditory processing can be observed in tests of pitch and duration, suggesting that her disorder is not specific to language. Linguistic analysis of signed, spoken, and written English indicates her language system is intact, but compromised because of impoverished input during the critical period for acquisition of spoken phonology. Specifically, although her sign language phonology is intact, spoken language phonology is markedly impaired. The authors argue that deprivation of auditory input during a period critical for the development of a phonological grammar and auditory–verbal short-term memory has limited her lexical and syntactic development in specific ways.

Journal

Brain and Language 1998; 63: 381–425

Neurocase Reference Number:

P568

Primary diagnosis of interest

Auditory agnosia, Landau–Kleffner syndrome, sign language

Author's designation of case

TM

Key theoretical issue

- Failure of patient to develop spoken English is due to distorted auditory input during critical period for auditory language acquisition, rather than damage to language system itself

Key words: auditory agnosia; Landau–Kleffner syndrome; aphasia

Scan, EEG and related measures

MRI, PET, EEG

Standardized assessment

WAIS, Raven Progressive Matrices, WMS-R, WRAT3, Finger Oscillation, Benton Facial Recognition Test, Trail Making Test, Wisconsin Card Sorting Test, Pyramids and Palm Trees Test, Boston Naming Test, PPVT-R

Other assessment

Audiometric screening, sound discrimination tests, speech discrimination tests, written grammar and comprehension tests

Lesion location

- PET: Bilateral temporal lobe

Lesion type

Hypometabolism

Language

English

Pure word deafness after bilateral primary auditory cortex infarcts

H. B. Coslett, H. R. Brashear and **K. M. Heilman**

Abstract

In pure word deafness, comprehension and repetition of speech are impaired, but reading, writing, and spontaneous speech are preserved. Pure word deafness is distinguished from generalized auditory agnosia by the preserved ability to recognize environmental sounds. The authors examined a patient with pure word deafness associated with bilateral infarctions of the primary auditory cortex, who could use auditory affective intonation to enhance comprehension. The primary auditory cortex seems to be essential for comprehending speech, but comprehension of non-verbal sounds and affective prosody may be mediated by other cerebral structures such as the auditory association cortex.

Journal

Neurology 1984; 34: 347–52

Neurocase Reference Number:

P569

Primary diagnosis of interest

Pure word deafness

Author's designation of case

Not mentioned

Key theoretical issue

- Preserved recognition of environmental sounds and affective intonation

Key words: pure word deafness; auditory agnosia; recognition of affective prosody

Scan, EEG and related measures

CT

Standardized assessment

Boston Diagnostic Aphasia Examination

Other assessment

Pure tone audiometry, competing environmental sounds, staggered spondaic words, synthetic sentence index, Ekman faces

Lesion location

- Enhancing infarct in the mid-portion at the left superior temporal gyrus involving Heschl's area plus an old infarct involving the middle and superior temporal gyri; the supramarginal gyrus; inferior portions of the postcentral, central, and precentral gyri; and the right inferolateral frontal lobe

Lesion type

CVA

Language

English

Pure word deafness and bilateral posterior perisylvian softenings: report of a case with neuropsychological-CAT correlation

M. Dalla Pria, H. Spinnler and **G. Vallar**

Abstract

A case of pure word deafness (PWD) with bilateral perisylvian softenings is described. The CAT findings fit the classical anatomical counterpart of PWD: a nearly complete acoustic disconnection of the Wernicke area. The associated severe apraxic disorders are discussed.

Journal

Archives Suisses de Neurologie, Neurochirurgie et de Psychiatrie 1979;125: 47–58

Neurocase Reference Number:

P570

Primary diagnosis of interest

Pure word deafness

Author's designation of case

Not mentioned

Key theoretical issue

- Neural basis of pure word deafness

Key words: pure word deafness

Scan, EEG and related measures

CAT, repeated technetium brain scans

Standardized assessment

Token Test, Raven's Coloured Matrices, Poppelreuter–Ghent's Overlapping Figures, Univariate Elementary Patterns, Farnsworth Test, Colour-Figure Matching Test

Other assessment

Sound discrimination tests, pure tone audiometry

Lesion location

- Bilateral hypodense area in the first temporal and supramarginal gyri and in the pre- and post-rolandic regions. On the right side, additional hypodense area in second and third gyri

Lesion type

Bilateral perisylvian softenings

Language

English

Evidence from a case of pure word deafness

G. Denes and **C. Semenza**

Abstract

In a patient with a classical syndrome of pure word deafness following a cerebrovascular accident, detailed neuropsychological examination showed an almost absolute inability to name meaningful non-verbal sounds, in spite of normal recognition as demonstrated by ability to match the sound with the corresponding natural source. This selective anomia, confined to auditory modality, is discussed in view of previous research in the general field of naming disturbances, which showed that for the vast majority of aphasics, anomia was not modality-dependent. Besides that, the contrasting performance of the patient, who showed complete lack of phonemic analysis matched with normal recognition of some acoustic parameters of speech such as prosody and stress, is discussed.

Journal

Cortex 1975; 11: 401–11

Neurocase Reference Number:

P571

Primary diagnosis of interest

Pure word deafness

Author's designation of case

RR

Key theoretical issue

- Comprehension but anomia for non-verbal sounds in a case of pure word deafness

Key words: pure word deafness; anomia; environmental sounds

Scan, EEG and related measures

EEG

Standardized assessment

WAIS, Token Test

Other assessment

Goldman perimetry, audiometric screening, sound localization, rhyming ability, musical sequence discrimination, timbre, pitch and loudness discrimination, young–old voice discrimination, foreign word discrimination, semantic hearing test, recognition of non-verbal sounds

Lesion location

- EEG abnormalities over left temporal lobe

Lesion type

Not mentioned

Language

English

Clinical and neuroradiological findings in a case of pure word deafness

M. Di Giovanni, G. D'Alessandro, S. Baldini, D. Cantalupi and **E. Bottacchi**

Abstract

Pure word deafness is a clinical syndrome included among the aphasias and is marked by complete deafness of sudden onset with conserved ability to understand and read the written word and with no speech disorders. The authors report the case of a 61- year-old man in whom pure word deafness developed after two episodes of acute cerebral ischaemia in quick succession. Neuroimaging procedures, that is: computed tomography scan, single photon emission computed tomography and magnetic resonance imaging, revealed the presence of two ischaemic lesions in the temporal cortex bilaterally. Neurophysiological investigations (electroencephalogram, brainstem auditory evoked potentials and stapedial reflex) were also studied. The authors discuss the outcome of all these investigations in the light of the relevant published work.

Journal

The Italian Journal of Neurological Sciences 1992; 13: 507–10

Neurocase Reference Number:

P572

Primary diagnosis of interest

Pure word deafness

Author's designation of case

Not mentioned

Key theoretical issue

- Neurophysiological and structural bases for pure word deafness

Key words: pure word deafness; temporal ischaemia; SPECT

Scan, EEG and related measures

CT, HMPAO-SPECT, EEG

Standardized assessment

Brainstem auditory evoked potentials

Lesion location

- Bilateral temporal lobe

Lesion type

Cerebral embolism

Language

English

The functional anatomy of recovery from auditory agnosia. A PET study of sound categorization in a neurological patient and normal controls

A. Engelien, D. Silbersweig, E. Stern, W. Huber, W. Doring, C. Frith and **R. S. Frackowiak**

Abstract

H2(15)O-PET was used to investigate the functional anatomy of recovery in a patient (J.B.) with bilateral perisylvian strokes and auditory agnosia, who partially regained the ability to recognize environmental sounds, but remained clinically word-deaf. The patient and a group of six normal volunteers were scanned in the following three conditions: (i) passive listening to environmental sounds; (ii) categorization of environmental sounds; (iii) at rest. In normal subjects, passive listening as compared with rest was associated with significant activations in the auditory cortices and posterior thalami, and in the inferior parietal lobe and anterior insula/frontal opercular region on the right. In J.B., activations were observed in the spared auditory cortex and inferior parietal lobe of the right hemisphere and in regions adjacent to the perisylvian lesion in the left hemisphere (anterior insula/frontal opercular region, middle temporal gyrus and inferior parietal lobe). The recovered function, as measured by categorization of sounds compared with passive listening, in J.B. was associated with bilateral activation of a distributed network comprising (pre)frontal, middle temporal and inferior parietal cortices, as well as the right cerebellum and the right caudate nucleus. In addition, there was a left-sided activation of the anterior cingulate gyrus. In normal subjects, the same categorization task led to activation of a network comprising (pre)frontal, middle temporal and inferior parietal cortices in the left hemisphere only. These results suggest that bilateral activation (with recruitment of areas homologous to those known to be responsible for normal function), the engagement of peri-infarct regions, and the involvement of a more widespread neocortical network, are mechanisms of functional reorganization after injury that may enable recovery from, or compensation for, cognitive deficits.

Journal

Brain 1995; 118: 1395–409

Neurocase Reference Number:

P573

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

JB

Key theoretical issue

- Functional anatomy of recovery in a case of auditory agnosia

Key words: auditory agnosia; recovery; audition; cognition; PET

Scan, EEG and related measures

PET, MRI, SPM maps

Standardized assessment

Leistungsprufsystem, Corsi Block Tapping, Nonverbaler Lerntest, Verbaler Lerntest, Aachen Aphasia Test

Other assessment

Audiometric screening, logical reasoning task, auditory tasks with environmental sounds, linguistic and paralinguistic auditory tasks, passive listening versus rest, categorization versus listening

Lesion location

- Left hemisphere: lesioning of third frontal gyrus, inferior parts of the pre- and post-central gyri, superior temporal gyrus, supramarginal gyrus, insula and deep basal ganglia. Primary auditory cortex completely destroyed. White matter underlying middle temporal gyrus and inferior parietal lobe partly destroyed
- Right hemisphere: small lesion of posterior part of the superior temporal gyrus, including part of the transverse temporal gyrus (Heschl's gyrus, primary auditory cortex) and posterior and ventral part of the insular cortex

Lesion type

CVA

Language

English

Dissociations in auditory word comprehension; evidence from 9 fluent aphasic patients

S. Franklin

Abstract
Traditional aphasia classifications do not allow for a detailed description of auditory comprehension impairments. A cognitive neuropsychological model of lexical processing allows us to distinguish at least five different levels of impairment in single word auditory comprehension. It also specifies a more complex relationship between impairments in repetition and auditory comprehension. Nine fluent aphasic patients, with auditory comprehension disorders, were accessed using tests of phoneme discrimination, lexical decision, synonym matching and word and non-word repetition. The results of these tests, as predicted, indicated that there were five dissociable levels of impairment, although there was some evidence for interaction between levels. The patients showed qualitative differences between auditory and written comprehension. Four of the patients had impairments in repetition despite having no impairment in phoneme discrimination tests. No two patients showed exactly the same pattern of impairment across all the tests administered.

Journal
Aphasiology 1989; 3: 189–207

Neurocase Reference Number:
P574

Primary diagnosis of interest
Forms of word deafness

Author's designation of case
ES, FM, NH, EC, AH, MK, DRB, CJ, DI

Key theoretical issue
- Clinical presentation of five different forms of word deafness

Key words: pure word deafness; word-form deafness; word meaning deafness; auditory agnosia; repetition

Scan, EEG and related measures
Not mentioned

Standardized assessment
Not mentioned

Other assessment
Phoneme discrimination, lexical decision, synonym matching, repetition

Lesion location
- Not mentioned

Lesion type
All CVA except AH – head inury

Language
English

Abstract word meaning deafness

S. Franklin, D. Howard and K. Patterson

Abstract
The authors report the case of a patient, DRB, who shows impaired auditory comprehension of abstract but not concrete words, in the context of preserved comprehension of all types of written words. Good performance on auditory lexical decision for abstract words indicates that DRB is able to access these lexical entries. This pattern may be termed 'abstract word meaning deafness'. Repetition is also more impaired for abstract words than concrete words. DRB is unable to repeat non-words or to write them to dictation. He makes semantic errors in real word repetition, which is compatible with the view that his repetition is semantically mediated. Exactly the same pattern of semantic errors and a particular difficulty with abstract words is found in tasks where DRB is required to write to dictation. DRB's auditory comprehension and repetition impairments are not affected by word frequency. Performance on specific words is inconsistent, and is improved following a written cue.

Journal
Cognitive Neuropsychology 1994; 11: 1–34

Neurocase Reference Number:
P575

Primary diagnosis of interest
Word meaning deafness

Author's designation of case
DRB

Key theoretical issue
- Clinical presentation of word meaning deafness

Key words: word meaning deafness; imageability; repetition

Scan, EEG and related measures
CT

Standardized assessment
Raven's Standard Progressive Matrices, British Picture Vocabulary Scale, Test for the Reception of Grammar

Other assessment
Pure tone audiometry, Pyramid and Palm Trees test, spoken and written word comprehension, phoneme discrimination, lexical decision, dictation, repetition

Lesion location
- Left middle cerebral infarct

Lesion type
CVA

Language
English

A distinctive case of word meaning deafness?

S. Franklin, J. Turner, M. A. Lambon Ralph, J. Morris and **P. J. Bailey**

Abstract

The authors report the case of a patient DrO, who has impaired auditory comprehension but intact written comprehension. His ability to repeat words that he cannot comprehend, and his good performance on auditory lexical decision, suggest that DrO has an impairment of the mappings between the lexical representations of spoken words and their corresponding semantic representations; a word meaning deafness. DrO's ability to understand heard words depends on their imageability and length, such that he is worse at comprehending words that are both short and abstract. This is interpreted in terms of phonological and semantic distinctiveness.

Journal

Cognitive Neuropsychology 1996; 13: 1139–62

Neurocase Reference Number:

P576

Primary diagnosis of interest

Word meaning deafness

Author's designation of case

DrO

Key theoretical issue

- Description of a disorder in which impaired comprehension of spoken words occurs without generalized semantic or phonological impairment

Key words: word meaning deafness; imageability

Scan, EEG and related measures

CT

Standardized assessment

Not mentioned

Other assessment

Pure tone audiometry, spoken and written word comprehension, sound and phoneme discrimination, repetition, lexical decision

Lesion location

- Left middle cerebral infarct

Lesion type

CVA

Language

English

Auditory sound agnosia without aphasia following a right temporal lobe lesion

T. Fujii, R. Fukatsu, S. Watabe, A. Ohnuma, K. Teramura, I. Kimura, S. Saso and **K. Kogure**

Abstract

A 55-year-old right-handed man showed inability to recognize the meaning of non-verbal sounds without impairment of language comprehension after a cerebrovascular accident. His auditory acuity was intact and no other sign of agnosia, apraxia or aphasia was detectable. His errors on a test of sound recognition were acoustic rather than semantic. Brain CT scan showed a small lesion in the posterior part of the right temporal lobe. This case suggests that auditory sound agnosia without language disorder can ensure a lesion confined to the right hemisphere, and that the deficit is discriminative rather than associative in nature.

Journal

Cortex 1990; 26: 263–8 Published erratum appears in Cortex 1990; 26: 672

Neurocase Reference Number:

P577

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Auditory agnosia for non-verbal sounds in context of preserved verbal comprehension may be associated with selective damage to right hemisphere

Key words: auditory agnosia; right hemisphere; aphasia

Scan, EEG and related measures

CT

Standardized assessment

Standard Language Test of Aphasia, WAIS, Seashore Measure of Musical Talents

Other assessment

Audiometric screening, recognition of non-verbal sounds

Lesion location

- Posterior part of right temporal lobe

Lesion type

Hyperdensity

Language

English

Pure word deafness and hemispheric dynamics: a case history

M. S. Gazzaniga, A. V. Glass, M. T. Sarno and **J. B. Posner**

Abstract

The authors report on the effects of a presumed unilateral brain lesion on language functions in a case of 'pure word deafness'. The patient was totally impaired in his auditory perception of speech while retaining much written comprehension, spontaneous speech and writing. As is usually the case, the word deafness was contaminated by some indication of auditory agnosia and some specific aphaisc deficits. The patient's performance on auditory–tactile matching tests provides a striking contrast with the performance of split brain patients. The right hemisphere in these patients is able to make both a visual–tactile and an auditory–tactile match between word and object. This case, however, was unable to make an auditory–tactile match. A possible explanation for the lack of right hemispheric language function in this case is offered. The damaged left hemisphere may have an interfering effect on the potential linguistic capacity of the non-domimant hemisphere.

Journal

Cortex 1973; 9: 136–43

Neurocase Reference Number:

P578

Primary diagnosis of interest

Pure word deafness

Author's designation of case

WB

Key theoretical issue

- Bilateral functional impairment from a unilateral left lesion producing pure word deafness

Key words: auditory agnosia; pure word deafness

Scan, EEG and related measures

Carotid arteriogram

Standardized assessment

Benton Sound Recognition Test, Benton and Spreen Neurosensory Center Comprehensive Examination for Aphasia, Token Test

Other assessment

Audiometric testing, speech discrimination, lateralization of sound, delayed auditory feedback, word discrimination, short-term memory

Lesion location

- Not mentioned

Lesion type

CVA

Language

English

Psychoacoustical deficits related to bilateral subcortical haemorrhages. A case with apperceptive auditory agnosia

O. Godefroy, D. Leys, A. Furby, J. DeReuck, C. Daems, P. Rondepierre, B. Debachy, J. Deleume and **A. Desaulty**

Abstract

The authors report a case of acute deafness secondary to bilateral haemorrhages involving the external capsule and extending to both temporal isthmi. The lesions probably disrupted both auditory radiations. Deafness disappeared within 2 weeks leading to a transient auditory agnosia for environmental and verbal sounds. Performance on audiological and neurolinguistic tests were consistent with the hypothesis of a deficit of non-specific auditory processes. Psychoacoustical deficits related to subcortical lesions are very rare and might differ from those due to cortical lesions by the lack of aphasia. The present case and both previous cases with subcortical lesions might suffer from auditory agnosia of apperceptive type. Its characteristics and the role of non-specific auditory processes are discussed using the data obtained from this third case.

Journal

Cortex 1995; 31: 149–59

Neurocase Reference Number:

P579

Primary diagnosis of interest

Apperceptive auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Auditory agnosia without aphasia can result from subcortical lesions sparing the temporal cortex

Key words: auditory agnosia; deafness; subcortex; apperceptive agnosia

Scan, EEG and related measures

CT, MRI, PET

Standardized assessment

MMSE, WAIS, PM38, Peterson Paradigm, Battery 144, Gestural Apraxia, Cancellation Test, Rey's Figure copying, Shortened Token Test, BDAE

Other assessment

Audiometric screening, brainstem auditory evoked potentials, sound recognition test, neurolinguistic assessment, oral comprehension, repetition, naming, fluency

Lesion location

- MRI: Bilateral external capsule extending to temporal isthmus

Lesion type

Haemorrhage

Language

English

A 66-year-old woman with a rapidly progressing dementia and basal ganglia involvement

M. Gold, A. Rojiani and **R. Murtaugh**

Abstract
A 66-year-old woman presented with a rapidly progressive dementia initially characterized by an auditory agnosia. She experienced a rapid progression of her aphasia and developed ataxia and myoclonus. An initial neurological evaluation suggested a left parieto-temporal lesion, however, neuroimaging did not reveal any. An MRI of her brain demonstrated highly focal T2 hyperintensities in her basal ganglia. The patient was diagnosed with Creutzfeldt–Jacob disease (CJD) on the basis of the presence of two proteins in the cerebrospinal fluid that are highly sensitive and specific for CJD. Pathological examination confirmed the diagnosis. The differential diagnosis and utility of MRI in patients with CJD is discussed.

Journal
Journal of Neuroimaging 1997; 7: 171–5

Neurocase Reference Number:
P580

Primary diagnosis of interest
Auditory agnosia; Creutzfeldt–Jacob disease

Author's designation of case
Not mentioned

Key theoretical issue
- Auditory agnosia may be a presenting feature of CJD

Key words: auditory agnosia; CJD; aphasia

Scan, EEG and related measures
MRI, EEG, lumbar puncture

Standardized assessment
None mentioned

Other assessment
Tests of verbal and visual memory, oral comprehension, repetition, reading, object naming, writing

Lesion location
- MRI: basal ganglia

Lesion type
Hyperintensity

Language
English

Auditory agnosia and cortical deafness: analysis of a case with 3-year follow-up

M. N. Goldstein, M. Brown and **J. Hollander**

Abstract
Studies of a patient with an auditory recognition disorder are presented. These are discussed in light of their relation with decreased auditory acuity. Auditory agnosia is not necessarily a static sign; improvement may occur. Auditory agnosia may not be a pure disturbance in cerebral associative function.

Journal
Brain and Language 1975; 2: 324–32

Neurocase Reference Number:
P581

Primary diagnosis of interest
Auditory agnosia

Author's designation of case
JS

Key theoretical issue
- Patient presented with generalized auditory agnosia which improved over time until speech comprehension was the only major deficit

Key words: auditory agnosia; pure word deafness

Scan, EEG and related measures
EEG

Standardized assessment
Porteus Maze, Bender Gestalt Test, Standard Achievement Test, Picture Interest Inventory, Differential Aptitude Tests, WAIS

Other assessment
Audiometric screening, speech sound discrimination, memory tasks, sound localization, pulse tone patterns reproduction, rhythmic stimuli reproduction

Lesion location
- Bilateral frontal and temporal regions, more pronounced on right

Lesion type
Possible necrosis secondary to encephalitis

Language
English

Mutism and auditory agnosia due to bilateral insular damage—role of the insula in human communication

M. Habib, G. Daquin, L. Milandre, M. L. Royere, M. Rey, A. Lanteri, G. Salamon and **R. Khalil**

Abstract

A case is reported of transient mutism and persistent auditory agnosia due to two successive ischaemic infarcts mainly involving the insular cortex on both hemispheres. During the 'mutic' period, which lasted about 1 month, the patient did not respond to any auditory stimuli and made no effort to communicate. On follow-up examinations, language competences had re-appeared almost intact, but a massive auditory agnosia for non-verbal sounds was observed. From close inspection of lesion site, as determined with brain resonance imaging, and from a study of auditory evoked potentials, it is concluded that bilateral insular damage was crucial to both expressive and receptive components of the syndrome. The role of the insula in verbal and non-verbal communication is discussed in the light of anatomical descriptions of the pattern of connectivity of the insular cortex.

Journal

Neuropsychologia 1995; 33: 327–39

Neurocase Reference Number:

P582

Primary diagnosis of interest

Mutism, auditory agnosia

Author's designation of case

TM

Key theoretical issue

- This patient's generalized auditory agnosia developed into an agnosia for non-verbal sounds only. The insula may have a critical role in attentional aspects of communication

Key words: speech; mutism; insular cortex; auditory agnosia

Scan, EEG and related measures

CT, MRI

Standardized assessment

BDAE, Rey–Osterrieth Complex Figure, Trail Making Test, Stroop Test, Wisconsin Card Sorting Test, Wechsler Memory Scale, Rey's 15 word auditory learning, WAIS-R

Other assessment

Audiometric screening

Lesion location

- Bilateral insular region

Lesion type

Ischaemic infarction

Language

English

A study of pure word-deafness

R. E. Hemphill and **E. Stengel**

Abstract

The authors describe a case in which there was a syndrome consisting of pure word deafness and what has been described as asymbolia for pain, following a head injury. The inter-relationship of these two disorders has been discussed and it has been pointed out that pure word deafness and asymbolia for pain have the common feature of a disturbance in the relationship between the individual and his environment. The localization of the responsible lesions has been discussed. Consideration has been given to the problem of the position of pure word deafness in the systems of the aphasias

Journal

Journal Neurology and Psychiatry 1940; 3: 251–62

Neurocase Reference Number:

P583

Primary diagnosis of interest

Pure word deafness

Author's designation of case

Henry G

Key theoretical issue

- Pure word deafness and its relationship to asymbolia for pain

Key words: pure word deafness

Scan, EEG and related measures

Not mentioned

Standardized assessment

Not mentioned

Other assessment

Audiometric screening

Lesion location

Not mentioned

Lesion type

Head injury

Language

English

Cortical auditory disorder caused by bilateral temporal infarctions

K. Ishii, Y. Ueda, N. Ohkoshi, H. Mizusawa and **S. Shoji**

Abstract

The authors present a 55-year-old man who suddenly became afflicted with a bilateral auditory disturbance. Auditory acuity was preserved relatively well. Brainstem auditory evoked potentials (BAEPs) were normal. Middle latency auditory evoked potentials (MLAEPs) revealed the total absence of all waves. Brain computed tomography (CT) showed infarctions of the temporal lobes bilaterally, involving the superior temporal and transverse temporal gyri. The recognition of verbal and non-verbal sounds was also impaired. Brain CT, MLAEPs and auditory recognition tests were useful in diagnosing the cortical auditory disorder.

Journal

Internal Medicine 1995; 34: 801–5

Neurocase Reference Number:

P584

Primary diagnosis of interest

Cortical auditory disorder

Author's designation of case

Not mentioned

Key theoretical issue

- Cortical auditory agnosia is a distinct syndrome from auditory agnosia and pure word deafness. Recognition of verbal and non-verbal stimuli are similarly impaired

Key words: auditory agnosia; deafness; evoked potentials; temporal lobe

Scan, EEG and related measures

CT, ECG

Standardized assessment

WAIS-R, WMS-R, Token Test, Western Aphasia Battery, audiometric screening

Other assessment

Brainstem auditory evoked responses, auditory recognition tests

Lesion location

- CT: bilateral temporal lobe (Heschl's and superior temporal gyri)

Lesion type

Infarct

Language

English

Defective auditory recognition after small haemorrhage in the inferior colliculi

K. Johkura, S. Matsumoto, O. Hasegawa and **Y. Kuroiwa**

Abstract

We report the case of a male patient with a traumatic small haemorrhage partially involving the bilateral inferior colliculi without evidence of a temporal lobe lesion. He was unable to comprehend spoken words although he had intact speech production, reading and writing abilities. Comprehension of environmental sounds was also affected. Among the receptive musical abilities, discrimination of intensity, tone and rhythm were preserved, while recognition of melody was impaired. Audiometry showed normal thresholds for pure tone. Waves I–IV of brainstem auditory evoked potentials were elicited normally, whereas the wave V was elicited with reduced amplitude and prolonged latencies on both sides. The main component of middle latency auditory evoked potentials, which is evoked over both hemispheres by monaural stimulation to either side in normal subjects, was elicited only over the hemisphere contralateral to the ear receiving stimulation. The patient's auditory findings were similar to those usually found in generalized auditory agnosia. Auditory agnosia is usually considered as a sign of a bitemporal cortical or subcortical disorder, but, in the patient, a brainstem disorder caused a disturbance of auditory recognition similar to auditory agnosia due to a bitemporal lesion. The patient's auditory findings may belong to the category of a brainstem auditory-processing disorder brought on by a small haemorrhage in the inferior colliculi. In addition, the impairment in the patient implies that, in the neural processing of musical parameters, the decoding of intensity, tone and rhythm is accomplished at the level of inferior colliculus, whereas further cortical processing is necessary for the appropriate recognition of melody.

Journal

Journal of the Neurological Sciences 1998; 161: 91–6

Neurocase Reference Number:

P585

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Case of generalized auditory agnosia resulting from brainstem pathology

Key words: auditory agnosia; inferior colliculus; word deafness; amusia

Scan, EEG and related measures

EEG, CT, MRI, HMPAO-SPECT

Standardized assessment

WAIS-R, Western Aphasia Battery, Standard Language Test of Aphasia

Other assessment

Audiometric screening

Lesion location

- Bilateral inferior colliculi

Lesion type

Haematoma

Language

English

Verbal auditory agnosia with focal EEG abnormality: an unusual case of a child presenting to an ENT surgeon with 'deafness'

U. Kale, M. El-Naggar and **M. Hawthorne**

Abstract

Verbal auditory agnosia implies the failure to recognize sounds in a patient who is nevertheless not deaf. The authors present a child referred to the ENT out-patient department for a suspected hearing loss investigation. At one point she had grommets inserted on the basis of a flat tympanogram, but with no effect. All hearing tests, including electrocochleography and distraction tests, revealed normal hearing thresholds. At the age of 3 years and 3 months, and on direct questioning of the parents, it was discovered that the child responded remarkably well to music. Paediatric neurologists made the diagnosis of verbal auditory agnosia. The condition is very similar to another acquired language disorder called Landau–Kleffner syndrome. It is unusual for such a case to present primarily to an ENT surgeon.

Journal

The Journal of Laryngology and Otology 1995; 109: 431–2

Neurocase Reference Number:

P586

Primary diagnosis of interest

Childhood verbal auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Verbal auditory agnosia in a case of possible Landau–Kleffner syndrome

Key words: agnosia; verbal; auditory; Landau–Kleffner syndrome

Scan, EEG and related measures

CT, EEG

Standardized assessment

Not mentioned

Other assessment

Pure tone audiometry

Lesion location

- No abnormalities noted

Lesion type

No abnormalities noted

Language

English

Subcortical auditory agnosia

S. Kazui, H. Naritomi, T. Sawada, N. Inoue and **J. I. Okuda**

Abstract

A case of generalized auditory agnosia without aphasia secondary to cardiogenic cerebral embolism is reported. The infarcts in this patient were localized within the bitemporal subcortices as confirmed by computerized axial tomography and magnetic resonance imaging. The findings suggested that interruption of both auditory radiations by bilateral subcortical lesions may play an important role in the occurrence of 'cerebral auditory disorders'.

Journal

Brain and Language 1990; 38: 476–87

Neurocase Reference Number:

P587

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Case of generalized auditory agnosia without aphasia due to small subcortical lesions

Key words: auditory agnosia; temporal lobe; subcortex

Scan, EEG and related measures

EEG, CT, MRI, cerebral angiography, electrocochleography

Standardized assessment

Standard language test of aphasia

Other assessment

Right–left discrimination, finger identification, colour recognition, visual object recognition, unilateral spatial neglect, constructional capacity, remote and recent memory, environmental sounds, verbal identification of melodies, loudness discrimination, frequency discrimination, click fusion tests, click detection threshold, discrimination lists with varying intervals, discrimination tests with varying number of clicks, vowel and phonemic discrimination – natural speech

Lesion location

- CT: right temporal and left parietal subcortex
- MRI: additionally left temporal stem

Lesion type

Infarction

Language

English

The problem of agnosia in the light of a case of pure word deafness

R. Klein and **J. Harper**

Abstract
A 45-year old man showed, after probable vascular lesion of the brain, a marked aphasic disorder and absence of response to auditory stimuli. After a short period lasting only a few days, the aphasic disturbance had almost completely disappeared; the picture which followed was that of a pure word deafness associated with disturbance in rhythm, musical expression and some disturbance in writing. On audiometric testing, carried out 6 weeks after the start of the illness, the air and bone conduction of the left ear was normal and there was a moderate mixed deafness in the right ear. At this period, the word deafness was still almost complete. The verbal sounds were experienced as an undifferentiated continuous humming noise without any rhythm. Subsequently a gradual limited improvement in the word sound perception occurred, but there was still a considerable defect after 18 months of observation. The sequence in which perception of various qualities of speech recovered was followed up, and a number of phenomena associated with the word deafness were studied. Some of these phenomena considered to be significant for the mechanism of the disorder have been discussed in detail.

Journal
Journal of Mental Science 1956; 102: 112–20

Neurocase Reference Number:
P588

Primary diagnosis of interest
Pure word deafness

Author's designation of case
RC

Key theoretical issue
- Nature of pure word deafness

Key words: pure word deafness; agnosia

Scan, EEG and related measures
Not mentioned

Standardized assessment
Terman–Merrill Vocabulary Tests

Other assessment
Audiometric screening, repetition tasks

Lesion location
- Not mentioned

Lesion type
CVA assumed

Language
English

Word-meaning deafness: a phonological–semantic dissociation

S. E. Kohn and **R. B. Friedman**

Abstract
Word-meaning deafness represents an auditory comprehension disturbance that is due to a dissociation between accurate phonological and semantic information. The patient is unable to understand a spoken word that he can repeat and can understand when read. Two cases of word-meaning deafness are presented to demonstrate that this disorder can be due either to a breakdown prior to the accessing of stored phonological representations ('pre-access') or to a breakdown following the accessing of stored phonological representations ('post-access').

Journal
Cognitive Neuropsychology 1986; 3: 291–308

Neurocase Reference Number:
P589

Primary diagnosis of interest
Word-meaning deafness

Author's designation of case
HN
LL

Key theoretical issue
- Word-meaning deafness represents either a deficit accessing representations in the phonological lexicon, or a disconnection between the phonological lexicon and semantic system

Key words: auditory agnosia; word-meaning deafness; phonological lexicon; semantic memory

Scan, EEG and related measures
CT

Standardized assessment
Boston Diagnostic Aphasia Examination

Other assessment
Tests of repetition, word–picture matching, word–body part matching, generation of definitions

Lesion location
- HN: Wernicke's area
- LL: Wernicke's area extending to body of left lateral ventricle

Lesion type
Not specified

Language
English

Auditory agnosia with relative sparing of speech perception

J. Lambert, F. Eustache, B. Lechevalier, Y. Rossa and **F. Viader**

Abstract

The authors report a case of auditory agnosia in which the initial clinical picture began with generalized auditory agnosia for verbal and non-verbal sounds, but rapidly changed to a selective auditory agnosia confined to the perception of non-verbal sounds. CT scanning and MRI did not demonstrate cortical or subcortical damage, except for bilateral ventricular enlargement. The patient was submitted to audiological investigations including physical and psychoacoustic studies. Deficits were revealed during the decay and loudness discrimination test, but no temporal auditory acuity deficit was observed. The results of these studies are discussed in relation to the clinical picture. Also, the dissociation between verbal perception and non-verbal perception is discussed.

Journal

Cortex 1989; 25: 71–82

Neurocase Reference Number:

P590

Primary diagnosis of interest

Auditory agnosia secondary to head injury

Author's designation of case

ID

Key theoretical issue

- Patient with generalized auditory agnosia developing to selective deficit for non-verbal sounds (although some evidence of difficulty with phonemic identification)

Key words: auditory agnosia; speech

Scan, EEG and related measures

CT, MRI, EEG

Standardized assessment

Rey Figure copy, French Aphasia Battery Test, Lusher's Test

Other assessment

Audiometric screening, sound matching, music reception, sound discrimination tests, voice, rhythm and language perception, click counting, verbal comprehension test, written word matching test

Lesion location

- CT: bilateral cerebral ventricles

Lesion type

Haemorrhage

Language

English

Self-monitoring behaviour in a case of severe auditory agnosia with aphasia

R. C. Marshall, B. Z. Rappaport and **L. Garcia-Bunuel**

Abstract

This case report describes an unusual combination of speech and language deficits secondary to bilateral infarctions in a 62-year-old woman. The patient was administered an extensive series of speech, language, and audiologic tests and was found to exhibit a fluent aphasia in which reading and writing were extremely well preserved in comparison to auditory comprehension and oral expression, and a severe auditory agnosia. In spite of her auditory processing deficits, the patient exhibited unexpected self-monitoring ability and the capacity to form acoustic images on visual tasks. The manner in which she corrected and attempted to correct her phonemic errors, while ignoring semantic errors, suggests that different mechanisms may underlie the monitoring of these errors.

Journal

Brain and Language 1985; 24: 297–313

Neurocase Reference Number:

P591

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Patient with generalized auditory agnosia and fluent aphasia who, despite the auditory agnosia, showed awareness of her speech production errors

Key words: auditory agnosia; fluent aphasia

Scan, EEG and related measures

EEG, CT

Standardized assessment

Word Intelligibility Picture Identification Test, Peabody Picture Vocabulary Test, Token Test, Auditory Comprehension Test for Sentences, Porch Index for Communication Ability, Boston Diagnostic Aphasia Examination, Minnesota Test for Differential Diagnosis of Aphasia, WAIS, Raven's Coloured Progressive Matrices, Rey–Osterrieth Battery

Other assessment

Audiometric testing, reading comprehension battery for aphasia, multilingual aphasia test

Lesion location

- CT: bilateral middle and superior temporal gyri, extending on left to inferior parietal lobe and insula

Lesion type

Hypodensity

Language

English

Cortical auditory disorders: clinical and psychoacoustic features

M. F. Mendez and **G. R. Geehan**

Abstract
The symptoms of two patients with bilateral cortical auditory lesions evolved from cortical deafness to other auditory syndromes: generalized auditory agnosia, amusia and/or pure word deafness, and a residual impairment of temporal sequencing. On investigation, both had dysacusis, absent middle latency evoked responses, acoustic errors in sound recognition and matching, inconsistent auditory behaviours, and similarly disturbed psychoacoustic discrimination tasks. These findings indicate that the different clinical syndromes caused by cortical auditory lesions form a spectrum of related auditory processing disorders. Differences between syndromes may depend on the degree of involvement of a primary cortical processing system, the more diffuse accessory system, and possibly the efferent auditory system.

Journal
Journal of Neurology, Neurosurgery and Psychiatry 1988; 51: 1–9

Neurocase Reference Number:
P592

Primary diagnosis of interest
Generalized auditory agnosia

Author's designation of case
Case 1 and Case 2

Key theoretical issue
- Psychoacoustic features of cortical auditory disorders

Key words: auditory agnosia; fluent aphasia

Scan, EEG and related measures
CT, echocardiography, brainstem auditory evoked response

Other assessment
Pure tone audiometry

Lesion location
- Case 1: bilateral haemorrhagic temporal infarcts bilaterally involving the superior temporal gyri and surrounding hypodensity from temporal tips to parietal lobes
- Case 2: right frontoparietotemporal infarct and left parietotemporal lesion

Lesion type
Embolism

Language
English

Word deafness mistaken for Alzheimer's disease: differential characteristics

M. F. Mendez and **S. Rosenberg**

Abstract
The inability to understand spoken words may result from speech comprehension disorders such as Wernicke's aphasia, hearing loss, and pure word deafness (PWD). However, clinicians may still mistake PWD, a rare cortical disorder with an isolated inability to understand speech, with Alzheimer's disease (AD). The correct differentiation of PWD from AD has implications for the prognosis and treatment of these patients. This report evaluated a patient with word deafness who was misdiagnosed as having AD. The misdiagnosis caused her much distress as well as a delay in initiating proper management. Additionally, this report characterizes the clinical features of PWD and compares them to the language changes of AD and of other disturbances with which PWD is often confused.

Journal
Journal of the American Geriatrics Society 1991; 39: 209–11

Neurocase Reference Number:
P593

Primary diagnosis of interest
Pure word deafness

Author's designation of case
Not mentioned

Key theoretical issue
- Differential diagnosis of pure word deafness from Alzheimer's disease

Key words: pure word deafness; Alzheimer's disease

Scan, EEG and related measures
CT

Standardized assessment
Not mentioned

Other assessment
Audiometric screening, aphasia screening

Lesion location
- Hypodensity deep in the medial aspect of the left temporal lobe

Lesion type
CVA assumed

Language
English

Analysis of word comprehension in a case of pure word deafness

M. N. Metz-Lutz and **E. Dahl**

Abstract
A case of pure word deafness due to a left temporal infarct is reported. The results of dichotic tests suggest that auditory verbal material may be processed in the right hemisphere. The inability to repeat nonsense words, the frequent semantic paraphasias in real-word repetition tasks, and the capacity to give a partial account of the meaning of a word that the patient cannot repeat show that despite the impairment of the phonological analysis, lexical semantic processing is possible. An attempt is made to demonstrate that the patient resorts to this semantic processing and that this reflects the linguistic competence of the right hemisphere.

Journal
Brain and Language 1984; 23: 13–25

Neurocase Reference Number:
P594

Primary diagnosis of interest
Pure word deafness and deep dysphasia

Author's designation of case
GL

Key theoretical issue
- Involvement of right hemisphere in repetition

Key words: pure word deafness; deep dysphasia

Scan, EEG and related measures
CT, EEG, brain stem evoked responses

Standardized assessment
BDAE

Other assessment
Audiometric testing, non-verbal and verbal auditory, repetition of words and non-words, dichotic listening studies, auditory comprehension

Lesion location
- Left temporal hypodensity spreading from cortex to the deep structures

Lesion type
CVA

Language
English

Deep dysphasia: an analogue of deep dyslexia in the auditory modality

F. Michel and **E. Andreewsky**

Abstract
A right-handed patient, with two left hemisphere lesions, a small one in the prefrontal lobe and a larger one in the temporal, presents an unusual syndrome: a massive deficit for oral language (expression and comprehension) contrasting with a fairly good preservation of written language (expression and comprehension). The processing of isolated words and sentences has been extensively tested with repetition and dictation tasks. The patient performs rather well with nouns, verbs, and adjectives, poorly with adverbs and function words, and completely fails with nonsense words. A remarkable feature of his repetition is the frequency of semantic paraphasias. Thus, this patient exhibits a behaviour rather similar to deep dyslexia, hence the possible label 'deep dysphasia'. The paper presents a 'pre-understanding' hypothesis to account for such behaviours.

Journal
Brain and Language 1983; 18: 212–23

Neurocase Reference Number:
P595

Primary diagnosis of interest
Deep dysphasia

Author's designation of case
Not stated

Key theoretical issue
- Description of deep dysphasia as a syndrome associated with semantic errors during repetition of spoken words and writing to dictation

Key words: deep dysphasia; temporal lobe; word deafness; aphasia

Scan, EEG and related measures
CT

Standardized assessment
None specified

Other assessment
IQ, memory, oral comprehension, repetition, dichotic listening, written comprehension, tachistoscopic reading

Lesion location
- Left anterior frontal lobe, left temporal lobe

Lesion type
Possible infarct

Language
English

Auditory agnosia. Analysis of a case with bilateral subcortical lesions

N. Motomura, A. Yamadori, E. Mori and **F. Tamaru**

Abstract

A case of typical auditory agnosia is reported. It is unique in that the clinical picture began with generalized auditory agnosia for verbal and non-verbal sounds and changed, over a period of two months, to a restricted auditory agnosia, confined to the recognition of non-verbal sounds. The lesions, confirmed by CT scanning and MRI, were subcortical bilaterally, without evidence of cortical damage, and have not previously been described. The results of audiological studies on temporal discrimination were similar to those described for bilateral temporal lobe lesions. The relationships between click intervals and the threshold to detect sound, and between numbers of clicks and the threshold, were also examined. The authors observed a tendency for the threshold to decline as the number of clicks was increased or the interval between two clicks was shortened. These phenomena are similar to temporal summation in cellular neurophysiology and the authors speculate that this phenomenon is an important factor in the auditory disturbances shown by the present case.

Journal

Brain 1986; 109: 379–91 Published erratum appears in Brain 1986; 109: 1322

Neurocase Reference Number:

P596

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Apparently earliest report of auditory agnosia changing from a generalized to a selective non-verbal deficit, associated with subcortical lesions

Key words: auditory agnosia; subcortical lesion

Scan, EEG and related measures

CT, MRI, EEG

Standardized assessment

WAIS, Token Test

Other assessment

Audiometric testing, environmental sounds, rhythm and melody recognition tests, vowel identification, phonemic discrimination, temporal discrimination, music testing, dichotic listening test, click fusion test, loudness discrimination, click detection threshold

Lesion location

- CT: left posterior thalamus, right posterior region

Lesion type

Left: hyperdensity
Right: hypodensity

Language

English

Clinical and anatomic findings in a case of auditory agnosia

D. R. Oppenheimer and **F. Newcombe**

Abstract

A case is reported of severe agnosia for verbal and non-verbal sounds in a setting of well-preserved intelligence and transient dysphasic symptoms. The lesions responsible for this disorder were bilateral cerebral infarcts, one of which had virtually destroyed the areas of Wernicke and Broca. The relation between lesions and disabilities is discussed and comparisons made with other published cases.

Journal

Archives of Neurology 1978; 35: 712–9

Neurocase Reference Number:

P597

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Discussion of pathological findings in a patient with auditory agnosia

Key words: auditory agnosia; word deafness

Scan, EEG and related measures

None mentioned

Standardized assessment

None mentioned

Other assessment

Audiometric screening, block design, cube counting, non-verbal span, word repetition, sound tests

Lesion location

- Left inferior frontal region and inferior parietal lobule, right superior temporal gyrus

Lesion type

Infarct

Language

English

Slowly progressive pure word deafness

M. Otsuki, Y. Soma, M. Sato, A. Homma and **S. Tsuji**

Abstract

Among the reports of primary progressive aphasia, there are few about patients who exhibited progressive pure word deafness with detailed auditory and radiological examination as well as neuropsychological assessment. The authors describe a 67-year-old right-handed man who exhibited slowly progressive pure word deafness over a period of 9 years without exhibiting any other cognitive or mental deterioration. Magnetic resonance imaging of his brain revealed generalized cortical atrophy, particularly in the left superior temporal region. Auditory examination revealed severe disability in discriminating each syllable or mora of Japanese words, despite adequate auditory acuity. He also showed impairment in temporal auditory discrimination assessed by the click fusion test and the click counting test. His ability to discriminate meaningful environmental sounds was mildly impaired. This paper discusses the pathophysiology of slowly progressive pure word deafness over a period of many years which was not complicated by other language or cognitive dysfunctions.

Journal

European Neurology 1998; 39: 135–40

Neurocase Reference Number:

P598

Primary diagnosis of interest

Progressive pure word deafness

Author's designation of case

Not mentioned

Key theoretical issue

- Progressive form of pure word deafness

Key words: auditory agnosia; primary progressive aphasia; pure word deafness

Scan, EEG and related measures

MRI, I-IMP SPECT, brain stem evoked potentials

Standardized assessment

Western Aphasia Battery (Japanese version), Token Test, WAIS-R, Raven Coloured Progressive Matrices, Benton Visual Memory Scale

Other assessment

Click fusion and click counting test, environmental sounds, pure tone audiometry

Lesion location

- Generalized atrophy most striking in the left superior temporal gyrus

Lesion type

Progressive atrophy

Language

English

Right unilateral auditory agnosia following left lenticular haemorrhage

F. Pasquier, D. Leys, M. Steinling, J. D. Guieu, H. Petit and **J. Cambier**

Abstract

A 33-year-old patient who had had left lenticular haemorrhage presented with an inability to understand oral language, with the right ear, and, in a less dramatic way, non-verbal sounds. This unilateral auditory agnosia was first associated with a right motor under-utilization and right motor, sensitive, visual and auditive extinctions. Speech discrimination scores were 100% with the left ear and 15% with the right ear, even less in dichotic conditions. Tonal audiogram, as well as early and late components of the auditory evoked potentials were normal. Cerebral regional perfusion and metabolism were impaired over the left parietotemporal area. There was severe hypoactivation of the left hemisphere with right monaural verbal stimulations. Rehabilitation consisting of non-specific attention tasks and repetitions of words reaching only the right ear was undertaken 15 months after the stroke. The oral language comprehension improved, as did the left hemisphere activation, and the extinction phenomena disappeared, except for the auditory one. The unilaterality of the auditory agnosia could be due, in part, to a peculiar physiological processing in this patient, such as poor performance of his right ipsilateral auditory pathway which could be improved with practice. A striatal lesion could induce a spatial hemi-inattention as reflected by the multimodal extinction in this case. Besides, a lack of selective activation for verbal stimulation of the left hemisphere is suggested.

Journal

Revue Neurologique (Paris) 1991; 147: 129–37

Neurocase Reference Number:

P599

Primary diagnosis of interest

Unilateral auditory agnosia

Author's designation of case

850681

Key theoretical issue

- Auditory agnosia for right ear only

Key words: unilateral auditory agnosia

Scan, EEG and related measures

MRI, CT, SPECT, auditory evoked potentials

Other assessment

Audiometric screening, speech discrimination

Lesion location

- Left posterior putamen and posterior and inferior white matter

Lesion type

Haemorrhage

Language

French

Correlation between EEG and auditory perceptual measures in auditory agnosia

P. S. Pearce and **H. Darwish**

Abstract

The authors describe a child who acquired a gradual sustained speech deficit for which no specific aetiology was found, and who had an associated epileptogenic abnormality on EEG. Assessment of auditory perceptual skills and receptive language confirmed that gradual parallel improvement occurred with the EEG after therapeutic anticonvulsant blood levels were obtained. Prospective trials of anticonvulsant drugs in conjunction with serial measures of central auditory abilities are necessary to establish their value in the management of the linguistic deficit in children with auditory agnosia.

Journal

Brain and Language 1984; 22: 41–8

Neurocase Reference Number:

P600

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

JA

Key theoretical issue

- Auditory agnosia associated with epileptogenic abnormality and recovery following anticonvulsant medication

Key words: auditory agnosia; EEG abnormality; anticonvulsant medication

Scan, EEG and related measures

EEG, CT

Standardized assessment

Peabody Picture Vocabulary Test, Reynell Verbal Comprehension Scale, pre-school language scale, Leiter International Performance Scale, Wechsler Intelligence Scale for Children-Revised

Other assessment

Discrimination of speech sounds, discrimination of environmental sounds, speech discrimination

Lesion location

- CT – normal

Lesion type

CT – normal

Language

English

Functional dissociations following bilateral lesions of auditory cortex

I. Peretz, R. Kolinsky, M. Tramo, R. Labrecque, C. Hublet, G. Demeurisse and **S. Belleville**

Abstract

Two cases of patients with bilateral lesions of the superior temporal cortex are described showing a number of dissociations in the auditory domain. In both patients, perception of speech and environmental sounds was normal while perception of tunes, prosody and voice was affected. Also, both patients showed impaired melodic, but not rhythmic processing. It is suggested that melody processing is mediated by specialized cortical subsystems instead of a more general purpose auditory architecture.

Journal

Brain 1994; 117: 1283–301

Neurocase Reference Number:

P601

Primary diagnosis of interest

Amusia, auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Melody processing could be mediated by specialized cortical systems instead of a more general-purpose architecture

Key words: auditory agnosia; amusia; auditory cortex

Scan, EEG and related measures

CT scans

Standardized assessment

Boston Diagnostic Aphasia Examination, Wechsler adult intelligence scale, Wechsler memory scale, Token test, Warrington recognition memory test

Other assessment

Language examination, recall and recognition of familiar melodies, lyric recognition, discrimination and recognition of unfamiliar tunes, recognition of environmental sounds and musical instruments, prosody recognition, voice recognition and discrimination, singing and related expressive behaviour

Lesion location

- Rostral auditory association cortex (both cases)

Lesion type

Case 1: ischaemic infarction
Case 2: aneurysmal subarachnoid haemorrhage

Language

English

A case of auditory agnosia

M. Reinhold

Abstract
A patient is described who suffered from a defective understanding of sounds including speech and music, although her hearing was not impaired. She had also lost the understanding of rhythm. Her auditory imagery was very defective. She was unable to think in terms of abstractions. Many symbols, such as those of arithmetic and grammar, were without meaning for her. She showed therefore a degree of acalculia. Her spelling was moderately defective, as was her grammar in written (but not in spoken) language. She was unable to perform two tasks simultaneously. She had, for a time, at the onset of her illness, suffered from auditory hallucinations. Expressive speech was very little disturbed. Reading was not impaired as regards the motor aspect, although comprehension of content, when dealing with abstract ideas, was defective. Writing was only slightly impaired – again not in the least as regards the motor aspect, but rather as regards punctuation, grammar and spelling. The patient showed no apraxia, no astereognosis, and no disorientation in space or time. She had little or no insight into her disabilities. She unconsciously used tactics of distraction to hide them. She preserved, in spite of them, a lively interest in current events, music, clothes, pen friends, embroidery, photography, reading and charitable works.

Journal
Brain 1950; 73: 203–23

Neurocase Reference Number:
P602

Primary diagnosis of interest
Auditory agnosia

Author's designation of case
Not mentioned

Key theoretical issue
- Early description of generalized auditory agnosia without aphasia

Key words: auditory agnosia; aphasia; amusia

Scan, EEG and related measures
None mentioned

Standardized assessment
None mentioned

Other assessment
Sound interpretation, auditory memory, insight, spontaneous speech, naming of objects and colours, reading, writing, calculation, visual memory, orientation in space and time, general knowledge, memory, praxis, conceptual thinking, auditory imagery and hallucinations

Lesion location
- Not stated

Lesion type
Not stated

Language
English

Pure word deafness and unilateral right temporo-parietal lesion: a case report

M. Roberts, P. Sandercock and E. Ghadiali

Abstract
The authors discuss a patient with an agnosia predominantly for spoken words who had a unilateral right hemisphere temporo-parietal lesion demonstrated by computed tomography.

Journal
Journal of Neurology, Neurosurgery and Psychiatry 1987; 50: 1708–9

Neurocase Reference Number:
P603

Primary diagnosis of interest
Pure word deafness

Author's designation of case
Not mentioned

Key theoretical issue
- Pure word deafness in a right-handed patient associated with a right temporo-parietal lesion

Key words: pure word deafness; lesion localization; crossed aphasia

Scan, EEG and related measures
CT

Standardized assessment
WAIS, Handedness Inventory

Lesion location
- Right parietal lesion extending inferiorly into the posterior temporal lobe

Lesion type
Infarct

Language
English

Clinical and audiological findings in a case of auditory agnosia

G. Rosati, P. De Bastiani, E. Paolino, S. Prosser, E. Arslan and **M. Artioli**

Abstract

A case is reported of severe agnosia for verbal and non-verbal sounds without associated aphasic disorder. A CT scan revealed bilateral, temporal lobe lesions from two ischaemic accidents that had occurred 9 months apart. The search for subtle deficits in the patient showed normal sensitivity to changes in the intensity and frequency of simple sounds; in contrast, his ability to discriminate sound duration and musical note sequences was severely impaired. The simultaneous recording of the whole auditory-evoked response pattern revealed no abnormality in the early components, which reflect the activation of the auditory nuclei and pathways of the brain stem. However, the middle and late components were delayed and slowed. These results and others in the literature suggest that the neocortex in man, as in other mammals, plays an essential role in the temporal aspects of hearing. Also, the two main ingredients commonly recognized in auditory agnosia, i.e. word deafness and the inability to interpret non-verbal sounds, are caused by the disruption of elementary, bilaterally represented cortical functions which start the processing of every kind of auditory information.

Journal

Journal of Neurology 1982; 227: 21–7

Neurocase Reference Number:

P604

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

MC

Key theoretical issue

- Bilateral cortical mechanisms may be involved early in the perception of verbal and non-verbal auditory stimuli

Key words: auditory agnosia; word deafness; auditory-evoked potentials; bitemporal lesions

Scan, EEG and related measures

CT, EEG

Standardized assessment

Token Test, WAIS, WMS

Other assessment

Audiometric screening, language test, test of non-verbal sounds

Lesion location

- Bilateral temporal lobe, left more than right

Lesion type

Ischaemia

Language

English

An analysis of speech perception in word deafness

E. M. Saffran, O. S. Marin and **G. H. Yeni-Komshian**

Abstract

A patient with a rather pure word deafness showed extreme suppression of right ear signals under dichotic conditions, suggesting that speech signals were being processed in the right hemisphere. Systematic errors in the identification and discrimination of natural and synthetic stop consonants further indicated that speech sounds were not being processed in the normal manner. Auditory comprehension improved considerably however, when the range of speech stimuli was limited by contextual constraints. Possible implications for the mechanism of word deafness are discussed.

Journal

Brain and Language 1976; 3: 209–28

Neurocase Reference Number:

P605

Primary diagnosis of interest

Pure word deafness

Author's designation of case

Not mentioned

Key theoretical issue

- Formal analysis of speech perception in pure word deafness

Key words: auditory agnosia; pure word deafness; speech perception

Scan, EEG and related measures

EEG, carotid angiogram

Standardized assessment

Seashore Measure of Musical Talents, Dichotic Names Test

Other assessment

Audiometric screening, speech sound discrimination and comprehension, repetition tasks

Lesion location

- Not mentioned

Lesion type

Not mentioned

Language

English

Speech discrimination and lip reading in patients with word deafness or auditory agnosia

M. Shindo, K. Kaga and **Y. Tanaka**

Abstract
The purpose of this study was to assess the ability of four patients with word deafness or auditory agnosia to discriminate speech by reading lips. The patients were studied using nonsense monosyllables to tests for speech discrimination, a lip reading test, the Token Test for auditory comprehension, and the Aphasia test. Our results show that patients with word deafness or auditory agnosia without aphasia can improve speech comprehension by reading lips in combination with listening, as compared with lip reading or listening alone. In conclusion, lip reading was shown to be useful for speech comprehension among these patients.

Journal
Brain and Language 1991; 40: 153–61

Neurocase Reference Number:
P606

Primary diagnosis of interest
Pure word deafness

Author's designation of case
Case 1, Case 2, Case 3, Case 4

Key theoretical issue
- Influence of lip reading on speech comprehension in pure word deafness

Key words: pure word deafness; auditory agnosia; lip reading

Scan, EEG and related measures
CT, auditory brain stem responses

Standardized assessment
Token Test

Other assessment
Audiometric screening, speech discrimination test for auditory perception, lip reading test, speech discrimination, lip reading ability

Lesion location
- Case 1: bilateral auditory radiation
- Case 2: bilateral temporal
- Case 3: bilateral temporal
- Case 4: left temporal

Lesion type
Case 1: Moyamoya disease
Case 2: subarachnoidal haemorrhage
Case 3: cerebral infarction
Case 4: cerebral bleeding

Language
English

Auditory agnosia without aphasia

O. Spreen, A. L. Benton and **R. W. Fincham**

Abstract
A 65-year-old patient, after a cerebrovascular accident, showed an inability to understand the meaning of common sounds, but retained the full understanding of language. No other agnostic defects were found. Repeated examinations over a period of eighteen months appear to confirm the finding of an isolated sound agnosia. Autopsy study showed a large defect involving the right frontal, temporal and parietal areas as well as the insula. No damage was demonstrated in the left hemisphere or the corpus callosum.

Journal
Archives of Neurology 1965; 13: 84–92

Neurocase Reference Number:
P607

Primary diagnosis of interest
Auditory agnosia without aphasia

Author's designation of case
Not mentioned

Key theoretical issue
- Auditory agnosia without aphasia may result from damage to right hemisphere structures only

Key words: auditory agnosia; aphasia; right hemisphere

Scan, EEG and related measures
EEG, ECG

Standardized assessment
WAIS

Other assessment
Audiometric screening, right–left orientation, finger localization, visual attention tests, short-term memory, speech tests, word fluency, arithmetic tests, reading and writing tasks, sound discrimination, pitch discrimination task

Lesion location
- Right frontal, temporal, inferior parietal regions, insula

Lesion type
Haematoma

Language
English

Childhood auditory agnosia

L. K. Stein and **F. K. Curry**

Abstract
The authors present the case of a 22-year-old female unable to understand speech, but with an apparently normal sensitivity for pure tones. At about the age of 10 hearing was much improved, and was demonstrated to be normal at the age of 20. Speech audiometry was not successful. Speech output resembled that of a severely deaf person. The authors diagnose an isolated form of 'auditory agnosia'.

Journal
Journal of Speech and Hearing Disorders 1968; 33: 361–70

Neurocase Reference Number:
P608

Primary diagnosis of interest
Childhood auditory agnosia

Author's designation of case
Not mentioned

Key theoretical issue
- Auditory agnosia in childhood

Key words: auditory agnosia

Scan, EEG and related measures
EEG

Standardized assessment
Randall's Island Test, WAIS, Raven Progressive Matrices, Rorschach, Wepman Auditory Discrimination Test, IPTA

Other assessment
Audiometric screening, right–left discrimination, finger identification, number recognition, auditory and perceptual discrimination tests

Lesion location
- Not known

Lesion type
Not known

Language
English

Pure word deafness due to left hemisphere damage

N. Takahashi, M. Kawamura, H. Shinotou, K. Hirayama, K. Kaga and **M. Shindo**

Abstract
The authors report the case of a 55-year-old right-handed man who presented with a long-lasting pure word deafness following left thalamic bleeding. There was no sign of aphasia. The auditory deficit was specific for language, while recognition of music and environmental sounds was normal. CT, MRI and PET examinations showed that the lesion was anatomically and functionally confined to the left cerebral hemisphere, mainly the white matter of the temporal and parietal lobes. Wernicke's area was largely preserved. It is proposed that pure word deafness was consequent to the isolation of Wernicke's area from incoming auditory information due to the interruption both of the association fibres from the right auditory area travelling across the corpus callosum and of the left auditory radiations.

Journal
Cortex 1992; 28: 295–303

Neurocase Reference Number:
P609

Primary diagnosis of interest
Pure word deafness

Author's designation of case
Not mentioned

Key theoretical issue
- Neural basis of pure word deafness

Key words: pure word deafness; auditory agnosia

Scan, EEG and related measures
CT, MRI, PET

Standardized assessment
WAIS, Western Aphasia Battery

Other assessment
Audiometric testing, speech, music and environmental sound recognition tests

Lesion location
- Left thalamus, posterior internal capsule and white matter of the left temporal lobe

Lesion type
Haematoma

Language
English

Pure word deafness following bilateral lesions. A psychophysical analysis

Y. Tanaka, A. Yamadori and **E. Mori**

Abstract

A 26-year-old woman suffered from pure word deafness associated with receptive and expressive amusia after cerebral infarcts bilaterally in the temporoparietal lobes. She did not comprehend spoken words yet production of speech, reading and writing appeared to be unaffected. A disturbance of temporal resolution was found in the auditory, visual and somatosensory modalities. The clinical defect was limited only to the auditory modality. The authors suggest that the auditory modality is more dependent upon a temporal resolution than other sensory modalities.

Journal

Brain 1987; 110: 381–403

Neurocase Reference Number:

P610

Primary diagnosis of interest

Pure word deafness, amusia, auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Auditory modality is more dependent upon a temporal resolution than other sensory modalities

Key words: amusia; auditory agnosia; pure word deafness

Scan, EEG and related measures

CT, MRI, EEG

Standardized assessment

Token Test, Standard Language Test of Aphasia, WAIS

Other assessment

Test for memory and calculations, audiological examination, linguistic studies, tests of musical capacity, neurophysiological investigation, neuroacoustic studies, discrimination tests in various sensory modalities

Lesion location

- Left: middle and posterior portion of the superior gyrus involving most of Heschl's gyrus and the geniculotemporal auditory radiation; supramarginal and angular gyri
- Right: middle and posterior portion of the superior temporal gyrus and in the parietal lobe involving the supramarginal and angular gyri; Heschl's gyrus and the auditory radiation

Lesion type

Cerebral infarct, generalized convulsions

Language

English

Imageability and category-specificity

L. K. Tyler and **H. E. Moss**

Abstract

In this paper, the authors report the case of DrO, a patient who has been described as having a selective problem understanding the meaning of abstract words in the auditory modality. This claim is tested by means of an on-line semantic priming task, comparing the automatic activation of semantic information in both the auditory and visual modalities. Although DrO showed priming for both abstract and concrete words in the visual modality, there was only priming for concrete words in the auditory modality. However, DrO's reaction times and errors in the auditory priming study suggested that he might have a generalized auditory processing impairment. The authors tested and confirmed this hypothesis in a series of further studies. They propose an account of why a general auditory processing impairment would affect abstract words more than concrete words by appealing to an auditory analogue of Plaut and Shallice's (Cognitive Neuropsychology 1993; 10: 377–500) computational model of deep dyslexia.

Journal

Cognitive Neuropsychology 1997; 14: 293–318

Neurocase Reference Number:

P611

Primary diagnosis of interest

Word meaning deafness

Author's designation of case

DrO

Key theoretical issue

- Does a generalized auditory processing impairment produce word meaning deafness?

Key words: word meaning deafness; imageability; auditory impairment

Scan, EEG and related measures

CT

Standardized assessment

Not mentioned

Other assessment

Pure tone audiogram, online priming of lexical decision (visual and auditory), phoneme discrimination, repetition

Lesion location

- Left middle cerebral infarct

Lesion type

CVA

Language

English

The syndrome of auditory agnosia. A case report and an attempt at neuropsychological qualification

G. Ulrich

Abstract

Proceeding from the neuropsychological examination of a patient with an exceptionally selective impairment of auditory gnosis of vascular origin, we make an attempt to analyse structurally the syndrome of auditory agnosia, a study of which has been neglected in comparison with analyses of visual agnosia. In our analysis we refer to 28 case reports, documented in the literature. Decisive syndromal points of view are: (a) there is an absence of significant audiological deficiencies; (b) errors of the acoustic–discriminative type prevail in auditory–visual matching tasks; (c) the gnostic deficit is modality-specific; (d) the same items are variably reproduced on repeated presentation; (e) there is marked fluctuation of performance; (f) there is exceptional irreversibility of the impairment; (g) amusia is a more or less obligatory accompanying phenomenon; (h) in cases of vascular origin there is always a history of repeated temporal lobe damage, this damage being predominantly in the form of bilateral lesions. This analysis indicates a close structural analogy to the syndrome of perceptive visual agnosia. The authors place great importance in the relationship between approaches and models of cognitive psychology [Neisser (Cognitive Psychology. New York: Appleton, 1967)] which they see as important for further theoretical development in neuropsychology.

Journal

Archiv für Psychiatrie und Nervenkrankheiten 1977; 224: 221–33

Neurocase Reference Number:

P612

Primary diagnosis of interest

Auditory agnosia

Author's designation of case

Not mentioned

Key theoretical issue

- Metanalysis of 28 case reports of auditory agnosia

Key words: pure word deafness; categorical processing; acoustic-discriminative errors; repeated temporal lobe impairment

Scan, EEG and related measures

Not mentioned

Standardized assessment

Not mentioned

Other assessment

Not mentioned

Lesion location

- Not mentioned

Lesion type

Not mentioned

Language

German

Clinical picture and morbid anatomy in a case of 'pure word deafness'

G. Wohlfart, A. Lindgren and B. Jernelius

Abstract

Pure word deafness is rare, but of great theoretical interest. The authors observed a patient in whom there was almost complete acoustic agnosia (verbal, musical and general) but no word blindness, paraphasia or agraphia. The internal language was intact. It was possible to examine the brain following the patient's death $1\frac{1}{2}$ years later. This paper reports in detail the clinical observations in this case and discuss these in the light of the cerebral lesions. Similar cases have been reported earlier as 'pure word deafness' although 'isolated acoustic agnosia' would perhaps be more descriptive and adequate.

Journal

Journal of Nervous and Mental Disease 1952; 116: 818–27

Neurocase Reference Number:

P613

Primary diagnosis of interest

Pure word deafness

Author's designation of case

Not mentioned

Key theoretical issue

- Clinical and post-mortem findings in a case of pure word deafness

Key words: auditory agnosia; amusia

Scan, EEG and related measures

Pneumoencephalogram, EEG, post-mortem examination

Other assessment

Audiometric screening, speech, music and environmental sound discrimination, voluntary speech, foreign language comprehension, writing and calculation tasks

Lesion location

- Large lesion in the region of the right Sylvian fissure, smaller lesions in the base of the right cerebellar hemisphere and left occipital lobe

Lesion type

Embolism

Language

English

Pure word deafness (acquired verbal auditory agnosia) in an Arabic speaking patient

B. A. Yaqub, G. G. Gascon, M. Al-Nosha and **H. Whitaker**

Abstract
A 38-year-old, right-handed Arabic-speaking male developed pure word deafness 3 days after myocardial infarction. He could recognize Arabic music and instruments but not words of songs; a radio broadcast from the Koran, but not the individual words; a male as opposed to female voice; Arabic and non-Arabic languages; and whether sentences were questions, exclamations, or imperatives. He discerned whether the speaker was emotionally neutral, happy, angry or sad. Contextual cues and reducing the rate of speaking aided verbal comprehension. Pure tone threshold audiometry revealed mild bilateral sensorineural loss up to 2000 Hz and a moderate high frequency loss. Brainstem auditory evoked potentials were normal, cortical auditory evoked potentials abnormal. CT scan revealed bilateral infarcts subcortically just posterior to the left superior temporal gyrus and the right posterior superior and midtemporal regions. Neurolinguistic tests indicated that the deficit is prephonemic and not due to impairment of linguistic discrimination.

Journal
Brain 1988; 111: 457–66

Neurocase Reference Number:
P614

Primary diagnosis of interest
Pure word deafness

Author's designation of case
Not mentioned

Key theoretical issue
- Clinical findings in a case of pure word deafness

Key words: pure word deafness; auditory agnosia

Scan, EEG and related measures
CT, EEG, brain stem evoked potentials, cortical auditory evoked potentials

Other assessment
Audiometric screening, verbal and non-verbal sound assessment, speech discrimination tasks, reading aloud and for comprehension, praxis, right–left discrimination, finger identification, naming of body parts, geometrical shapes and colours, simple addition, copy figure, immediate and remote memory, click fusion and click counting, vowel identification, phonemic discrimination/identification

Lesion location
- Bilateral temporal regions

Lesion type
Infarct

Language
English

Word deafness and Wernicke's aphasia: report of cases and discussion of the syndrome

D. K. Ziegler

Abstract
A case of complete auditory agnosia ('pure word deafness') and one of Wernicke's aphasia are reported. The difference between the two syndromes is discussed. Total deafness and auditory hallucinations occurred at different times in the course of the illness of the patients with auditory agnosia. Their significance is commented on. Severe behavioural disturbance occurred in both patients. The difficulty of its differentiation from purely psychogenic disease is indicated, and its relation to temporal lobe dysfunction is suggested.

Journal
Archives of Neurology and Psychiatry, Chicago 1951; 67: 323–31

Neurocase Reference Number:
P615

Primary diagnosis of interest
Pure word deafness and Wernicke's aphasia

Author's designation of case
Case 1: CN
Case 2: BI

Key theoretical issue
- Dissociation of pure word deafness from generalized aphasia

Key words: pure word deafness; auditory agnosia; Wernicke's aphasia

Scan, EEG and related measures
EEG, pneumoencephalogram

Standardized assessment
Goldstein Stick Test

Other assessment
Case 1: spontaneous speech, reading, writing, naming, calculation, comprehension of auditory stimuli, comprehension of environmental sounds, music interpretation
Case 2: spontaneous speech and behaviour, repetition, recognition of written and spoken words, writing, calculation, abstraction tests, naming

Lesion location
- Not known

Lesion type
Not known

Language
English