PREVIOUS CASES

Pure Alexia

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Pure alexia is a reading disorder that occurs in literate
dividuals secondary to a lesion in the left occipito-temporal
region. The hallmark of this deficit is the word-length effect:
the naming latencies of patients increase dramatically with
increasing numbers of letters in the word. For example, every
additional letter can take a further 3 s for a patient who is
reading a word aloud [compared with 9 ms for a normal age-
matched reader; Behrmann et al., 1998 (P728)]. Pure alexia
is also known as letter-by-letter reading, spelling dyslexia,
alexia without agraphia, verbal dyslexia, word blindness or
letter-by-letter dyslexia.

Pure alexia was first extensively described by Déjerine.
This reading pathology is a key syndrome in the ‘language
zone’ theory of the French neurologist. In his book entitled
‘Sémiologie des affections du système nerveux’ (1914),
Déjerine describes pure alexia as a disconnection syndrome
that isolates the ‘center for the optic images of letters’,
situated in the left angular gyrus, from both visual cortices.
Because this language center cannot be accessed through
visual stimulation, the patients cannot read. However, they
are still able to write, speak and understand speech because
the lesion is peripheral, i.e. out of the language zone per se.
Alexia is said to be ‘pure’ because it is not associated with
any other language disorders.

Following Déjerine’s work, many neurologists have
attempted to locate the exact reading pathway in the human
brain [see Black and Behrmann (1994) for a review]. They
were either interested in the minimal pathway necessary for
reading, or in the potential existence of a common pathway
for reading and visual object perception or color perception
[Woods and Pöppel, 1974 (P774); Michel et al., 1979
(P760); Chanoine et al., 1998 (P736)]. The account that the
neurologists have defended or tried to falsify is the hypothesis
of a disconnection between the visual areas and the language
zone (the left angular gyrus) being responsible for pure alexia
[Déjerine, 1892 (P739); Déjerine and Viallet, 1893 (P740);
Greenblatt, 1973, 1976, 1990 (P744–746); Stachowiak and
Poeck, 1976 (P771); Vincent et al., 1977 (P773); Yamadori,
1980 (P775); Binder and Mohr, 1992 (P733); Hickok et al.,
1995 (P749); Beversdorf et al., 1997 (P732)]. Another critical
question concerns the role of a lesion in the splenium of the
corpus callosum in giving rise to pure alexia [Fox and
Hillemand, 1925 (P743); Ajax et al., 1977 (P725); Kurachi
et al., 1979 (P756); Quint and Gilmore, 1992].

Pure alexia has also generated a considerable interest in
the field of cognitive neuropsychology. The general approach
in this field is to isolate the cognitive components (potentially
damaged in the patients) that are necessary for normal
reading. Several issues have been addressed in that direction.

One issue concerns the functional locus of the deficit that
is responsible for pure alexia. Three potential loci (at least)
have been proposed. Pure alexia is attributed to a low-level
perceptual deficit [Kinsbourne and Warrington, 1963 (P755);
Levine and Calvania, 1978 (P757); Patterson and Kay, 1982;
Friedman and Alexander, 1984; Farah and Wallace, 1991;
Rapp and Caramazza, 1991; Hanley and Kay, 1992;
Behrmann and Shallice, 1994; Perri et al., 1996 (P767);
Sekuler and Behrmann, 1996 (P769); Behrmann et al., 1998
(P728); Chialant and Caramazza, 1998 (P737); Miozzo and
Caramazza, 1998 (P761)], a deficit in letter recognition
[Levine and Calvania, 1978 (P757); Reuter-Lorenz and
Brann, 1990; Howard, 1991; Kay and Hanley, 1991 (P754);
Arguin and Bub, 1993; Behrmann and Shallice, 1995; Bub
and Arguin, 1995; Behrmann et al., 1998 (P728); Saffran
and Coslett, 1998], or a more central impairment at (or after)
the level of the orthographic lexicon, sometimes referred to
as ‘the visual word form system’ [Déjerine, 1892 (P739);
Warrington and Shallice, 1980; Katz, 1990 (P753); Schacter
et al., 1990 (P768); Hanley and Kay, 1996 (P748); Arguin
et al., 1998 (P726)].

Another issue that is currently under investigation concerns
the residual reading abilities, and in particular the lexical
effects, that are observed in many pure alexic patients [see
Behrmann et al. (1998; P728) for a review]. These abilities
can be due to the residual functioning of the system that supported reading pre-morbidly [Bub and Arguin, 1995; Behrmann et al., 1998 (P756); Behrmann, 1998 (P728); Montant et al., 1998 (P764); Behrmann et al., 2000 (P730)]. Alternatively, they could be due to a compensatory system that supports reading after brain damage [Shallice and Saffran, 1986; Coslett and Saffran, 1989, 1994; Coslett et al., 1993; Feinberg et al., 1995 (P742); Buxbaum and Coslett, 1996 (P734); Saffran and Coslett, 1998]. According to the latter view, letter-by-letter (overt) reading is the product of the left hemisphere, while lexical effects derive from covert reading, which is supported by the right hemisphere. This hypothesis is controversial, in particular because pure alexic patients do not show the pattern of semantic errors that is typically associated with right hemisphere language processing.

The other issues that are addressed in pure alexia concern recovery and rehabilitation methods [Kurachi et al., 1979 (P756); Behrmann et al., 1990; Daniel et al., 1992; Arguin and Bub, 1994; Behrmann and McLeod, 1995 (P727); Seki et al., 1995 (P76); Maher et al., 1998 (P759); Nitzberg et al., 1999 (P765)], letter-by-letter reading as a compensatory strategy [Speedie et al., 1982 (P770); Kay and Hanley, 1991 (P754); Price and Humphreys, 1992; Greenwald and Gonzalez-Rothi, 1998 (P747)], the associated syndromes, like music alexia or number alexia [Caplan and Hedley-Whyte, 1974 (P735); Cohen and Dehaene, 1995 (P738); Horikoshi et al., 1997 (P750)], and the cross-linguistic variability [Karanth, 1981 (P752); Mochizuki and Ohtomo, 1988 (P762); Sugishita et al., 1992 (P772); el Alaoui-Faris et al., 1994 (P741)].

References


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Alexia without agraphia and the inferior splenium

E. Ajax, T. Schenkenberg and M. Kostejanetz

Abstract
A patient who had alexia without agraphia, right homonymous hemianopia, and intact color naming was studied anatomically. Pathologic involvement of the splenium and related forceps was restricted to the inferior third, supporting published suggestions that inferior elements of this commissure and left peristriate cortex may be essential to the decoding to the written word, while colour-naming may be functionally aligned to more dorsal elements.

Journal
Neurology 1977; 27: 685–8

Neurocase Reference Number:
P725

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
Mr. A

Key theoretical issue
• The process of decoding written words depends on the inferior calcarine cortex, splenium and related forceps. The dorsal fibres of the splenium may be ‘sufficient’ for colour naming

Key words: pure alexia; inferior splenium; reading pathway

Scan, EEG and related measures
EEG, arteriogram

Standardized assessment
Halstead–Wepman Aphasia Screening Test, Gray’s Oral Reading Test, WAIS, Graham–Kendall Memory-for-Designs Test

Other assessment
Nil

Lesion location
• Left occipital lobe, inferior one-third of the forceps major and contiguous splenium of the corpus callosum. Entire left lingual, the cuneus, the inferior lip of the precuneus and the posterior half of the fusiform gyrus. Extensive destruction of the subjacent white matter in all of these areas

Lesion type
Infarction

Language
English

Extent and limits of covert lexical activation in letter-by-letter reading

M. Arguin, D. Bub and J. Bowers

Abstract
The occurrence of implicit reading in brain-damaged patients with letter-by-letter dyslexia suggests a process of covert lexical activation, whereby lexical access occurs on the basis of parallel letter encoding. The extent and limitations of this process were studied by examining masked orthographic and phonological word priming as well as orthographic neighbourhood size effects in letter-by-letter reader IH. In Experiment 1, masked repetition priming occurred with primes displayed in case-alternated format that were shown for 100 ms (a duration that does not reliably support overt word identification in IH). Under similar exposure conditions, however, primes that are homophones to the target failed to affect performance, in contrast to neurologically intact observers (Experiment 2). Experiment 3 showed that IH’s naming latencies are reduced for words with many (versus few) orthographic neighbours. This result suggests that overt word recognition in the patient is not strictly mediated by sequential letter recognition, but rather that it is conjointly affected by covert lexical activation. Relative to neurologically intact subjects, however, the pattern of the neighbourhood size effect shown by IH as a function of word frequency is abnormal and suggests that lexical activation based on the parallel processing of letters is weakened in the patient compared with normal readers. Overall, results from IH point to a weak form of covert lexical activation phenomena that accompany the disorder.

Journal

Neurocase Reference Number:
P726

Primary diagnosis of interest
Letter-by-letter reading, pure alexia

Author’s designation of case
IH [also described in Bowers et al., 1996a,b (Cognitive Neuropsychology 1996; 13: 415–42; 525–67)]

Key theoretical issue
• Word recognition in pure alexia is not strictly mediated by sequential letter identification but is affected by (weakened) covert lexical activation

Key words: letter-by-letter reading; covert reading; implicit reading

Scan, EEG and related measures
None

Standardized assessment
Nil

Other assessment
Nil

Lesion location
• Haematoma in the left temporo-occipital area

Lesion type
Subarachnoid haemorrhage that was drained surgically

Language
English
Rehabilitation for pure alexia: efficacy of therapy and implications for models of normal word recognition

M. Behrmann and J. McLeod

Abstract
In an attempt to remediate the deficit in letter processing thought to underlie pure alexia or letter-by-letter reading, a therapy programme was implemented with SI, a 46-year-old woman with pure alexia following a posterior cerebral artery infarction. Prior to the intervention programme, SI's reading and letter processing abilities were characterized in detail, and baseline measurements were taken to rule out spontaneous recovery or fluctuation in her performance. A 9-week intensive therapy programme was designed to alter SI's report of single letters as a function of serial position in a string from the observed linear function to a more normal M-shaped curve. This involved training SI to apprehend and report the identity of the letter in the final position of a string as string length was increased and exposure duration decreased. Whereas SI's ability to recognize the final letter improved significantly on post-therapy measures, there was no significant transfer to her word reading performance. The word length effect—increase in reaction time with increasing word length—remained unchanged from pre- to post-therapy, although the intercept shifted downwards. These results raised questions about the relationship between processes involved in single letter report and word reading in normal processing. Findings from rehabilitation studies such as this are not only useful in determining the efficacy of particular intervention techniques but, importantly, shed light on the mechanisms underlying normal cognitive behaviour.

Journal
Neuropsychological Rehabilitation 1995; 5: 149–80

Neurocase Reference Number:
P727

Primary diagnosis of interest
Pure alexia

Author's designation of case
SI (same patient as Behrmann et al., Cognitive Neuropsychology 1998; 15: 7–51, Behrmann et al., Neuropsychologia 1998; 36: 1115–32; note they used the initials IS for the same patient)

Key theoretical issue
• Although patient SI improved in her ability to report single letters following therapy, this improvement did not alter her letter-by-letter reading ability

Key words: pure alexia; single letter report; rehabilitation

Scan, EEG and related measures
MRI

Standardized assessment
Boston Naming Test, Reading Comprehension Battery for Aphasia, Western Aphasia Battery, Wechsler Memory Scale

Other assessment
Nil

Lesion location
• Infarction of posterior cerebral artery resulting in left temporal–occipital lesion and involving the hippocampus, fusiform and lingual gyri; smaller lesion affecting the parietal region also noted

Lesion type
Infarction

Language
English

Visual complexity in letter-by-letter reading: ‘pure’ alexia is not pure

M. Behrmann, J. Nelson and E. B. Sekuler

Abstract
Standard accounts of pure alexia have favoured the view that this acquired disorder of reading arises from damage to a left posterior occipital cortex mechanism dedicated to the processing of alphanumerical symbols. We challenge these accounts in two experiments and demonstrate that patients with this reading deficit are also impaired at object identification. In the first experiment, we show that a single subject, EL, who shows all the hallmark features of pure alexia, is impaired at picture identification across a large set of stimuli. As the visual complexity of pictures increases, so EL’s reaction time to identify the stimuli increases disproportionately relative to the control subjects. In the second experiment, we confirm these findings with a larger group of five pure alexic patients using a selected subset of high- and low-visual complexity pictures. These findings suggest that the deficit giving rise to pure alexia is not restricted to orthographic symbols per se but, rather, is a consequence of damage to a more general-purpose visual processing mechanism.

Journal
Neuropsychologia 1998; 36: 1115–32

Neurocase Reference Number:
P728

Primary diagnosis of interest
Pure alexia

Author’s designation of case
Case 1: EL (same patient as Montant and Behrmann, Cognitive Neuropsychology 17: 2000: in press; P763)
Case 3: MW [same patient as Sekuler and Behrmann 1996; 13: 941–74; P769], Behrmann et al., Cognitive Neuropsychology 1998; 15: 7–51; (P729)]
Case 4: DK (same patient as Behrmann et al., Cognitive Neuropsychology 1998; 15: 7–51; P729)
Case 6: IS

Key theoretical issue
• Because the object identification ability of patients with pure alexia is influenced by the visual complexity of the stimulus to a greater extent than normal subjects, they appear not to have a pure reading deficit

Key words: pure alexia; word-meaning deafness; multiple sclerosis

Scan, EEG and related measures
Case 1: CT, MRI, angiogram
Case 2: CT
Case 3: MRI
Case 4: MRI
Case 5: MRI, EEG, SPECT
Case 6: MRI

Standardized assessment
Case 1: Visual Object and Space Perception Battery, PALPA, Spelling test, Factor referenced cognitive tests [Ekstrom et al. (Manual kit of factor-referenced cognitive tests, Princeton, NJ, USA: Educational Testing Service)], Snodgrass and Vanderwart pictures
Cases 2–6: Snodgrass and Vanderwart pictures

Other assessment
Nil
Lesion location
- Case 1: Left occipital and temporal cortices including peristriate inferotemporal association cortex and lateral posterolateral temporal neocortex
- Case 2: Left occipital lobe
- Case 3: Left occipital lobe
- Case 4: Left occipital lobe
- Case 5: No focal lesion; bilateral frontal slowing on EEG. Mildly decreased cerebral perfusion bilaterally on SPECT
- Case 6: Left occipitotemporal region including hippocampus, fusiform and lingual gyri; smaller lesion affecting the parietal region

Lesion type
Cases 1, 2, 3, 4 and 6: Infarction
Case 5: Closed head injury

Language
English

A literature review and new data supporting an interactive account of letter-by-letter reading

M. Behrmann, D. Plaut and J. Nelson

Abstract
We present a theoretical account of letter-by-letter (LBL) reading that reconciles discrepant findings associated with this form of acquired dyslexia. We claim that LBL reading is caused by a deficit that affects the normal activation of the orthographic representation of the stimulus. In spite of this lower-level deficit, the degraded orthographic information may be processed further, and lexical, semantic and higher-order orthographic information may still influence the reading patterns of these patients. In support of our position, we present a review of 57 published cases of LBL reading in which we demonstrate that a peripheral deficit was evident in almost all of the patients and that, simultaneously, strong effects of lexical/semantic variables were observed on reading performance. We then go on to report findings from an empirical analysis of seven LBL readers in whom we document the joint effects of lexical variables (word frequency and imageability) and word length on naming latency. We argue that the reading performance of these patients reflects the residual functioning of the same interactive system that supported normal reading pre-morbidly.

Journal
Cognitive Neuropsychology 1998; 15: 7–51

Neurocase Reference Number:
P729

Primary diagnosis of interest
Pure alexia

Author’s designation of case
Case 2: PC
Case 4: DK (same patient as Behrmann et al., Neuropsychologia 1998; 36: 1115–32)
Case 5: MS [same patient as Sekuler and Behrmann, Cognitive Neuropsychology 1996; 13: 941–74 (P769), Behrmann et al., Neuropsychologia 1998; 36: 1115–32 (P728), note they used the initials MA for the same patient]
Case 5: IS [same patient as Behrmann and McLeod, Neuropsychological Rehabilitation 1995 (P727); 5: 149–80 but reversed initials, Behrmann et al., Neuropsychologia 1998; 36: 1115–32 (P728)]

Key theoretical issue
- The data from the meta-analysis and from the new empirical studies support the view that the basic deficit giving rise to pure alexia is a peripheral deficit but that, through top-down connections, lexical and semantic knowledge can influence the reading performance of the patients

Key words: pure alexia; interactive account; peripheral deficit; lexical/semantic influences

Scan, EEG and related measures
Case 1: CT
Case 2: MRI
Case 3: MRI
Case 4: MRI
Case 5: MRI, EEG
Case 6: MRI
Case 7: MRI
Standardized assessment
Nil

Other assessment
Nil

Lesion location
- Case 1: Left occipital lobe
- Case 2: Left occipitotemporal
- Case 3: Left occipital lobe
- Case 4: Left occipital lobe
- Case 5: No focal lesion; bilateral frontal slowing on EEG
- Case 6: Left occipitotemporal region including hippocampus, fusiform and lingual gyri; smaller lesion affecting the parietal region
- Case 7: Left occipital and temporal lobe

Lesion type
Cases 1, 3, 4 and 6: Infarction
Case 2: Resected meningioma
Case 5: Closed head injury
Case 7: Resected arteriovenous malformation

Language
English

M. Behrmann, S. Shomstein, S. E. Black and J. J. S. Barton, in preparation

Neurocase Reference Number:
P730
Varieties of alexia. Word and letter blindness

D. F. Benson, J. Brown and B. Tomlinson

Abstract
Six case reports are presented, including three individuals with dominant medial occipital pathology (alexia without agraphia syndrome) and three with dominant frontal lesions (Broca’s aphasia). It was readily noted that the three patients with posterior lesions could read letters much better than words (word-blind) whereas the patients with anterior lesions could read words better than letters (letter-blind). Several explanations to account for this dichotomy are postulated.

Journal

Neurocase Reference Number:
P731

Primary diagnosis of interest
Alexia without agraphia, Broca aphasia

Author’s designation of case
Cases 1–3 (+ three Broca aphasic patients, Cases 4–6)

Key theoretical issue
- Alexia is not a uniform, monolithic entity. The verbal and literal alexia subtypes do exist clinically and indicate significant variations in the site of pathology

Key words: verbal alexia; literal alexia; word and letter blindness

Scan, EEG and related measures
Cases 1–3: Radioisotope brain scan

Standardized assessment
Case 1: Nil
Case 2: Koh’s blocks, Ishihara charts
Case 3: Babcock sentence

Other assessment
Nil

Lesion location
- Cases 1–3: Dominant (left) medial occipital region

Lesion type
Cases 1–3: Infarction due to occlusion of the left posterior cerebral artery

Language
English

Pure alexia: clinical–pathologic evidence for a lateralized visual language association cortex


Abstract
Traditional views of pure alexia have held that the disorder results from a disconnection between the secondary visual cortices of both hemispheres and the angular gyrus of the dominant hemisphere. Evidence has accumulated, however, suggesting the importance of the posterior inferior temporal area in visual language processing. We describe clinical–pathological support for the presence of a lateralized visual language association area in the dominant posterior inferior temporal lobe.

Journal
Clinical Neuropathology 1997; 16: 328–31

Neurocase Reference Number:
P732

Primary diagnosis of interest
Pure alexia

Author’s designation of case
Nil (same patient as Vincent et al., Neurology 1977; 27: 689–91; P773)

Key theoretical issue
- Pure alexia need not arise from disconnection of two hemispheres as traditionally argued; fusiform and/or lingual gyrus of dominant hemisphere involved in visual language processing

Key words: pure alexia; fusiform gyrus; lingual gyrus; disconnection

Scan, EEG and related measures
CT, post-mortem

Standardized assessment
Nil

Other assessment
Nil

Lesion location
- Posterior inferior surface of left temporal lobe with some compression of anteriormost occipital cortex; post-mortem revealed loss of grey matter in left fusiform and lateral lingual gyr, adjacent subcortical white matter and left occipital cortex

Lesion type
Meningioma resected

Language
English
The topography of callosal reading pathways. A case control analysis

J. R. Binder and J. P. Mohr

Abstract
Lesion topography and reading ability were analysed in 17 patients with dominant posterior cerebral artery territory infarction. Patients with dominant posterior cerebral artery infarction in whom reading was unaffected served as an anatomical control group. Normal readers had lesions in the medial and ventral occipital lobe, sparing dorsal white matter pathways and the ventral temporal lobe. Global and permanent alexia occurred only with additional injury to the splenium, forceps major or white matter above the occipital horn of the lateral ventricle. These data suggest that callosal pathways mediating reading lie above the occipital horn and have little connection with the ventromedial occipital region. Patients with ‘spelling dyslexia’ had large lesions of the ventral temporal lobe involving cortical regions believed to participate in later stages of visual processing. These findings provide a framework for the prediction of dyslexia type and severity based on lesion topography.

Journal
Brain 1992; 115: 1807–26

Neurocase Reference Number:
P733

Primary diagnosis of interest
Spelling dyslexia, global alexia

Author’s designation of case
Patient No 6
Patient No 7
Patient No 8
Patient No 9
Patient No 10
(five global alexic patients are also described)

Key theoretical issue
- The parallel streams participating in reading are distributed widely over the lateral and central surface of the occipito-temporal region, which explains the incomplete and qualitatively distinct reading disorders. The type and severity of reading deficit can be predicted from lesion site and volume

Key words: spelling alexia; global alexia; lesion topography

Scan, EEG and related measures
Cases 6–10: CT

Standardized assessment
Cases 6–10: Gray Oral Reading Test, Peabody Picture Vocabulary Test

Other assessment
Nil

Lesion location
- Cases 6–10: Calcarine cortex and underlying white matter of the medial occipital lobe, as well as the deep white matter of the occipito-temporal junction inferior to the occipital horn of the lateral ventricle. Cortex and white matter in the inferior temporal and anterior fusiform gyri

Lesion type
Cases 6–10: Infarction of the left posterior cerebral artery territory

Language
English

Deep dyslexic phenomena in a letter-by-letter reader

L. J. Buxbaum and H. B. Coslett

Abstract
Numerous accounts of pure alexia have suggested that a pre-lexical impairment precludes rapid access to orthographic information in patients with the disorder. We report a patient with features of both pure and partially recovered deep dyslexia in whom we demonstrate pre-lexical deficits in maintaining a reliable abstract representation of the right side of letter arrays, as well as in modulating a ‘spotlight’ of visual attention. These deficits, we suggest, encourage the patient’s use of a letter-by-letter reading strategy; despite them, however, he demonstrates rapid, accurate reading of some, but not all classes of words. Furthermore, the patient’s reading is influenced by both pre-lexical and lexical–semantic factors such that speed and accuracy are optimal for high imageability nouns of few letters. Finally, the patient accurately names orally spelled words of all classes. Taken together, these data are consistent with the hypothesis that rapid reading may be enabled by lexical–semantic support from a right hemisphere-mediated processing system which recognizes words as whole, thereby mitigating the effect of the pre-lexical deficits.

Journal
Brain and Language 1996; 54: 136–67

Neurocase Reference Number:
P734

Primary diagnosis of interest
Pure alexia, deep dyslexia

Author’s designation of case
JH

Key theoretical issue
- JH suffers from a pre-lexical deficit that forces him to derive lexical information through two routes: a rapid, whole word route which is reliant upon a right hemisphere lexicon, and a default letter-by-letter strategy

Key words: pure alexia; implicit reading; right hemisphere reading

Scan, EEG and related measures
MRI

Standardized assessment
Boston Diagnostic Aphasia Examination

Other assessment
Nil

Lesion location
- Left inferior temporo-occipital cortex, parahippocampal gyrus, left lateral geniculate

Lesion type
Infarction

Language
English
Cuing and memory dysfunction in alexia without agraphia. A case report

L. R. Caplan and T. Hedley-Whyte

Abstract
A 68-year-old female with rheumatic heart disease suddenly developed a right hemianopsia, right hemisensory loss, and complete alexia. An embolic occlusion of the left posterior cerebral artery had caused a large infarction involving the left temporal and occipital lobes and their subjacent white matter. A small infarction was seen in the left thalamus. Unusual features of this case included faulty number naming, disordered visual object and picture identification, and a severe defect in recent memory function which lasted 6 months. Emphasis is placed on the patient’s capabilities with visual symbols when naming was not required, and on her behaviour when cuing or prompting was introduced. The defect in alexia without agraphia may involve faulty visual exploration of a written word symbol, and not merely a pure disconnection between visual input in the right hemisphere and the language region of the left hemisphere.

Journal
Brain 1974; 97: 251–62

Neurocase Reference Number:
P735

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
Nil

Key theoretical issue
- The disorder in pure alexia is a modality specific visual disturbance. The defect in this syndrome may be one of faulty visual tuning. The fault may be helped by cuing. The left calcarine or paracalcarine cortex, or the thalamus may take part in tuning the visual analyser

Key words: alexia without agraphia; memory deficit; cuing

Scan, EEG and related measures
EEG, autopsy

Standardized assessment
Nil

Other assessment
Nil

Lesion location
- From the mid-portion of the left temporal lobe into the occipital lobe around the calcarine fissure, including the white matter, splenium, posterior hippocampus and geniculate bodies. Part of the posteroventral lateral nucleus of the left thalamus

Lesion type
Infarction following embolic occlusion

Language
English

Optic aphasia with pure alexia: a mild form of visual associative agnosia? A case study

V. Chanoine, C. Teixeira Ferreira, J. F. Demonet, J. F. Nespoulous and M. Poncet

Abstract
A single-case study is reported of a naming disorder selective to the visual modality. The patient showed intact access to structural knowledge of objects and letters, but impaired access to complete semantic knowledge of objects and alphabetical knowledge of letters from visual input. The impairment was most striking when the patient had to discriminate between semantically similar objects or within a given symbolic repertoire, i.e. letters. The co-occurrence of a partial deficit of visual recognition for objects and for letters indicated features of optic aphasia and pure alexia. This symmetric performance between object and letter processing may also constitute a mild form of visual associative agnosia.

Journal
Cortex 1998; 34: 437–48

Neurocase Reference Number:
P736

Primary diagnosis of interest
Optic aphasia, pure alexia, associative agnosia

Author’s designation of case
CN

Key theoretical issue
- Optic aphasia with pure alexia can be considered as the manifestation of a mild form of associative agnosia

Key words: optic aphasia; pure alexia; associative agnosia

Scan, EEG and related measures
MRI

Standardized assessment

Other assessment
See Howard and Orchard-Lisle (Cognitive Neuropsychology 1983; 1: 163–90) for semantic assessment

Lesion location
- Left lingual, fusiform and parahippocampal gyri, posterior portion of the internal capsule and thalamus on the left side, paracallosal lesion, complete interruption of the intra-hemispheric part of callosal fibres

Lesion type
Stroke in the territory of the left cerebral and left anterior choroidal arteries

Language
English
Perceptual and lexical factors in a case of letter-by-letter reading

D. Chialant and A. Caramazza

Abstract
We report the case of a letter-by-letter reader (MJ) who showed normal processing of single letters and who could normally access the orthographic input lexicon when presented with letter names for aural recognition, or when allowed enough time to process a visually presented letter string. However, MJ showed severe difficulties in simultaneously processing multiple letters and other simple visual stimuli. Furthermore, she does not have normal access to lexical orthographic representations and their meanings when stimuli are presented for too brief a time to allow serial processing of the letter string. We found no evidence of (partial) lexical or semantic access without corresponding recognition of the letters in a word. No signs of implicit reading were observed when the input stimuli were controlled for the relevant visual features; ‘implicit reading’ was only obtained under conditions that allowed sophisticated guessing. This pattern of results is interpreted as indicating that LBL reading (at least in MJ) results from damage to pre-lexical processing mechanisms. In MJ’s case, the deficit reflects the degraded transfer of information from a normal visual processing system in the right hemisphere to a normal language processing system in the left hemisphere.

Journal

Neurocase Reference Number:
P737

Primary diagnosis of interest
Letter-by-letter reading, pure alexia

Author’s designation of case
MJ

Key theoretical issue
- Pure alexia arises from a deficit that affects the transfer of intact visual information from the right hemisphere to a normal language processing system in the left hemisphere. The transmission of information from the right to the left hemisphere is serial, letter by letter

Key words: letter-by-letter reading; pure alexia; pre-lexical deficit; sequential letter identification

Scan, EEG and related measures
MRI

Standardized assessment
Nil

Other assessment
Nil

Lesion location
- 1.5 cm lesion in the left optic radiation near the thalamus long with multiple periventricular white matter lesions with involvement of the corpus callosum

Lesion type
Left occipital CVA

Language
English

Reading numbers in pure alexia: effects of task and hemispheric specialization

L. Cohen and S. Dehaene

Abstract
Selective conservation of the ability to read Arabic numbers in patients unable to read words or even letters is a classical characteristic of pure alexia described by Déjerine (1892). We report our work on the capacity of two patients with pure typical alexia to process numbers. Our main finding was that these patients could count pairs of Arabic numbers correctly when the reading task was simple (example 2 → four) or when the task involved comparing sizes (example 2 → four is bigger than two). However, these patients often made mistakes when asked to perform arithmetic operations (example 2 → two plus four equals six”). Using these two numbers, there was a similar dissociation between excellent performance on comparison tests and severe deficiency in reading out loud. We interpret these findings with the hypothesis that both of the hemispheres can identify Arabic numbers, but that the visual systems on the right and left play a different role during different tasks. In pure alexia, a lesion in the left identification system leads to selective deficiency in linguistic tasks such as reading numbers out loud, recognizing numbers with several figures or mental arithmetic. The right identification system in intact and is sufficient for comparison or reading isolated Arabic numbers.

Journal

Neurocase Reference Number:
P738

Primary diagnosis of interest
Pure alexia

Author’s designation of case
Case 1: GOD
Case 2: SMA

Key theoretical issue
- Because of their left hemisphere lesion, pure alexic patients are poor in processing visually presented numbers when the task involves linguistic operations. They use their intact right hemisphere to process numbers in non-linguistic tasks

Key words: pure alexia; number reading; hemispheric specialization

Scan, EEG and related measures
Nil

Standardized assessment
Cases 1 and 2: Boston Diagnostic Aphasic Examination

Other assessment
Nil

Lesion location
- Case 1: Infero-medial occipito-temporal lobe in the left hemisphere
- Case 2: Infero-medial occipito-temporal lobe in the left hemisphere, left thalamus

Lesion type
Cases 1 and 2: Infarction

Language
French
Contribution à l’étude anatomopathologique et clinique des différentes variétés de cécité verbale

J. Déjerine

Abstract
Nous devons admettre aujourd’hui en clinique, deux variétés bien distinctes de cécité verbale, relevant l’une et l’autre d’une localisation bien différente. La première variété est produite par une lésion siégeant dans la sphère du langage (pli courbe gauche); la deuxième, par une lésion siégeant dans la sphère visuelle commune séparant le pli courbe de cette dernière. Dans la première variété—cécité verbale avec agraphie ou altérations très marquées de l’écriture—les images optiques des lettres sont détruites et la cécité verbale s’accompagne soit d’agraphie totale, soit d’altérations très marquées des différents modes de l’écriture. Cette lésion intéressant la sphère du langage, il est facile de comprendre pourquoi les malades présentent, en général, un certain degré de paraphasie. La destruction du centre optique des lettres explique encore pourquoi on ne peut, chez eux, réveiller les images optiques des lettres à l’aide du sens musculaire. Dans la deuxième variété—cécité verbale pure avec intégrité de l’écriture spontanée et sous dictée—le centre des images optiques des lettres—pli courbe—est intact, mais la lésion le sépare, l’isole du centre visuel commun. Le pli courbe ne peut plus, par conséquent, être mis en jeu par une excitation visuelle. Par contre, il peut l’être: par une excitation volontaire, telle que l’écriture spontanée; —par une excitation auditive telle que l’écriture sous dictée; —ou encore par le sens musculaire (lettres tracées en l’air par les mains ou les pieds). L’écriture spontanée et sous dictée sont donc intactes; l’acte de copier, seul, est défectueux. La lésion étant ici complètement en dehors de la sphère du langage, les malades ne présentent pas de troubles de la parole, et leur langage intérior est intact. A l’aide des données précédentes, il sera toujours facile, en clinique, de reconnaître l’une ou l’autre de ces formes, qui correspondent, ainsi que je viens de l’établir, à des localisations bien distinctes.

Journal
Compte Rendu des Séances de la Société de Biologie 1892; 44: 61–90

Neurocase Reference Number:
P739

Primary diagnosis of interest
Word blindness (cécité verbale)

Author’s designation of case
C...(also described in Landolt, 1888, Book for Donders’ Jubilee)

Key theoretical issue
- Two varieties of word blindness, with distinct brain lesions, exist. The first variety is due to a lesion in the language sphere that destroys the ‘centre for the optic images of letters’ (located in the angular gyrus) and affects both the reading and writing abilities. Because the lesion is in the language sphere, word blindness is associated with some degree of paraphasia. The second variety (pure alexia) is due to a disconnection of the visual areas from the left angular gyrus. The centre for the optic images of letters is intact but isolated from the visual areas. The angular gyrus cannot be accessed through visual stimulation, which explains the reading impairment, but it can be activated during spontaneous writing, writing under dictation or kinesthetic writing. Because the lesion is outside the language areas, the patients have no language disorder associated with alexia

Key words: pure alexia; alexia with agraphia; disconnection; angular gyrus; optic images of letters

Scan, EEG and related measures
Autopsy

Standardized assessment
Snellen’s Scale

Other assessment
Nil

Lesion location
- First infarct: left lingual, fusiform gyri, cuneus, tip of the occipital lobe and splenium of the corpus callosum. Second infarct: left inferior parietal lobe and angular gyrus

Lesion type
Two infarctions (4 years apart)

Language
French
Contribution à l’étude de la localisation anatomique de la cécité verbale pure

J. Déjerine and N. Vialet

Abstract
Not available

Journal
Comptes Rendus des Séances de la Société de Biologie, 1893; 45: 790–3

Neurocase Reference Number: P740

Primary diagnosis of interest
Pure word blindness

Author’s designation of case
‘Le malade’ (same as in Déjerine, Soc Société de Biologie 1891; p. 197; Mémoires de la Société de Biologie, 27 février 1892)

Key theoretical issue
● The inferior part of the Burdach inferior longitudinal tract in the left hemisphere contains physiologically differentiated fibres that link together the visual zone and the language zone. In the patient, one pathway only of this tract was destroyed: the reading pathway

Key words: pure word blindness; lesion site

Scan, EEG and related measures
Autopsy, histologic exam (Pal method)

Standardized assessment
Nil

Other assessment
Nil

Lesion location
● Left lingual, fusiform, cuneus gyri and the tip of the occipital lob; see article

Lesion type
Infarction

Language
French

Alexia without agraphia in Arabic language. Neurolinguistic and MRI study

M. el Alaoui-Faris, F. Benbelaid, C. Alaoui, L. Tahiri, M. Jiddane, A. Amarti and T. Chkili

Abstract
A 33-year-old woman developed an alexia without agraphia, a colour anomia, a right hemianopia, an aphasic amnesia and a verbal amnesia. The brain MRI showed the lesions in the left splenium of corpus callosum, forceps major, optic radiations and anterior temporal lobe. The fact that she measured writing comprehension and had complete recovery of reading impairment despite the persistence of anatomic lesions plead in favor of an active participation of the right hemisphere (RH) on reading; this capacity of the RH may be due to the linguistic particularities of Arabic writing.

Journal

Neurocase Reference Number: P741

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
Case 3852/90

Key theoretical issue
● The right hemisphere seems to play an important role in the preservation and recovery of reading in Arabic speaking pure alexic patients, possibly because there is a strong visuo-spatial component in the processing of written Arabic language

Key words: alexia without agraphia; Arabic language; recovery

Scan, EEG and related measures
MRI, CT, EEG, angiography

Standardized assessment
Ishihara, Popelreuter’s pictures, Rey, Wechsler, Memory Battery of Signoret

Other assessment

Lesion location
● Left splenium of the corpus callosum, forceps major, optic radiations and anterior temporal lobe

Lesion type
Infarction

Language
French
Knowledge, implicit knowledge and metaknowledge in visual agnosia and pure alexia

T. E. Feinberg, D. Dyckes-Berke, C. R. Miner and D. M. Roane

Abstract
Residual or implicit knowledge has been observed in patients with object agnosia, optic aphasia and pure alexia. Previous investigators have considered implicit knowledge in these patients to be dissociated from awareness on the basis of intact semantic capabilities that are consistent with right hemisphere processing. The absence of explicit verbal identification is presumably dependent upon damaged left hemisphere systems. We describe a 72-year-old woman with a left occipital infarction, object agnosia and pure alexia who was unable to explicitly identify visual stimuli (objects and words), but was able to make reliable judgements of her residual knowledge on forced-choice matching tasks. While the patient could not consistently demonstrate awareness of knowledge prior to stimulus matching (‘Do you know what this is?’), she was able to reliably demonstrate awareness of knowledge for response accuracy (‘Are you sure?’) assessed after stimulus matching. Further, the extent of the patient’s metaknowledge corresponded to her degree of preserved knowledge. We propose that this pattern of performance suggests limited or partial access to preserved semantic knowledge which, though degraded, is not ‘non-conscious’.

Journal
Brain 1995; 118: 789–800

Neurocase Reference Number:
P742

Primary diagnosis of interest
Pure alexia, visual agnosia

Author’s designation of case
RE (same as Case 3 in Feinberg et al., Cortex 1994; 30: 395–411)

Key theoretical issue
- Implicit reading in pure alexia is not necessarily non-conscious, as can be demonstrated by the study of patient’s awareness of their perceptual ability

Key words: implicit knowledge; metaknowledge; associative visual agnosia; pure alexia

Scan, EEG and related measures
CAT scan

Standardized assessment
Western Aphasia Battery, Wechsler, The Assessment of Aphasia and Related Disorders, Benton Facial Recognition Test, Guide to the Standard Progressive Matrices, Osterrieth’s figures

Other assessment
Nil

Lesion location
- Total destruction of the calcarine cortex and extensive infarction of fusiform, lingual and parahippocampal gyri, the temporal white matter is extensively involved, the splenium of the corpus callosum is clearly involved

Lesion type
Complete posterior cerebral artery occlusion

Language
English

Rôle vraisemblable du splénium dans la pathologie de l’alexie pure par lésion de la cérébrale postérieure

C. Fox and P. Hillemand

Abstract
Not available

Journal

Neurocase Reference Number:
P743

Primary diagnosis of interest
Pure alexia

Author’s designation of case
Monsieur P.

Key theoretical issue
- The lesion of the splenium of the corpus callosum interrupts the transfer of the visual information from the intact right hemisphere to the language zone in the left hemisphere

Key words: pure alexia; splenium of the corpus callosum

Scan, EEG and related measures
Autopsy

Standardized assessment
Nil

Other assessment
Nil

Lesion location
- Territory of the left posterior cerebral artery (see article for a very precise description of the lesion sites)

Language
French
Alexia without agraphia or hemianopsia. Anatomical analysis of an autopsied case

S. H. Greenblatt

Abstract
This report describes the clinical and pathological findings of a woman who had a left occipital glioblastoma causing the syndrome of alexia without agraphia or hemianopsia. She died 6 days after operation and study of the brain confirmed the presence of an intact primary visual system on the left. The tumor had involved only the inferior outflow tracts of the left primary visual cortex (transverse fasciculus of the lingual gyrus and vertical occipital fasciculus) and the splenium of the corpus callosum. This mechanism for the production of the syndrome of alexia without agraphia is consistent with the disconnection theory which is usually adduced to explain the syndrome, but the entire superior outflow of the calcarine cortex and associated dorsal occipital cortex was intact. Thus, the syndrome of alexia without agraphia has been produced by a lesion which did not completely disconnect the intact left visual system from all its occipital association cortex. This fact has implications for the anatomical locations of the neural pathways for reading and for visual–verbal colour naming. It is proposed that, within each occipital lobe, the inferior association tracts and the ventro-medial (lingual and fusiform) gyri are necessary for reading. Visual–verbal colour naming, on the other hand, apparently may be served either by the dorsal or by the ventral outflow paths from calcarine cortex.

Journal
Brain 1973; 96: 307–16

Neurocase Reference Number:
P744

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
No. 274372-2

Key theoretical issue
● Complete disconnection of the left visual system from the occipital association cortex is not necessary for the occurrence of pure alexia. Damage to the inferior association tracts and the ventro-medial (lingual and fusiform) gyri is sufficient to impair reading

Key words: alexia without agraphia; disconnection; reading pathway

Scan, EEG and related measures
EEG, technetium 99 brain scan, arteriography, autopsy

Standardized assessment
Nil

Other assessment
Nil

Lesion location
● Inferomedial white matter of the left occipital lobe and splenium of the corpus callosum

Lesion type
Necrosis due to a glioblastoma multiforme

Language
English

Subangular alexia without agraphia or hemianopsia

S. H. Greenblatt

Abstract
A 40-year-old right-handed woman underwent resection of an unruptured vascular malformation which was located just deep to the left posterior insula. Postoperatively, she demonstrated a transient syndrome of alexia without or hemianopsia. Analysis of her postoperative findings, in conjunction with all available anatomical data, led to the conclusion that the responsible lesion was in the white matter of the left occipitotemporal region, below the angular gyrus and lateral to the lateral ventricle. Hence, the lesion in the present case was truly subangular and is therefore distinctly differentiated from the classical lesion in alexia without agraphia, which may be described as splenio-occipital.

Journal
Brain and Language 1976; 3: 229–45

Neurocase Reference Number:
P745

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
No. 154127-5

Key theoretical issue
● Pure alexia in this patient is ascribed to the incision of the white matter located inferior and deep to the left angular gyrus

Key words: alexia without agraphia; subangular lesion

Scan, EEG and related measures
EEG, arteriography

Standardized assessment
WAIS, reading Comprehension Subtest of the Peabody, Farnsworth–Munsell 100-Hue Test, AO H-R-R Pseudo-isochromatic Plates

Other assessment
Nil

Lesion location
● Cortical incision near the temporo-occipital junction, in or close to Brodmann’s area 37. Subjacent white matter incision more posteriorly and upward, closer to the white matter beneath the angular gyrus

Lesion type
Excision of a vascular malformation

Language
English
Left occipital lobectomy and the preangular anatomy of reading

S. H. Greenblatt

Abstract
Two patients had alexia after left occipital lobectomies. Case 1 was a 55-year-old man with a glioblastoma. At 4 months after surgery he could read slowly, but reading was neither efficient nor pleasant. Case 2 was a 19-year-old male who had a more restricted, medial occipital lobectomy for an encapsulated mesenchymal chondrosarcoma. The tumor did not invade brain initially, and the patient recovered efficient reading after 15 months. It is postulated that Case 2 was able to recover efficient reading because he still had a field of left occipitotemporal cortex connected to homologous cortex on the right.

Journal
Brain and Language 1990; 38: 576–95

Neurocase Reference Number:
P746

Primary diagnosis of interest
Alexia

Author’s designation of case
Case 1
Case 2

Key theoretical issue
• The known brain areas that support reading are the left pre-angular white matter and the left angulotemporal region of the cortex

Key words: alexia; lobectomy; reading pathway

Scan, EEG and related measures
Case 1: CT, arteriography
Case 2: CT, angiography

Standardized assessment
Case 1: Nil
Case 2: Wechsler, WRAT (Wide Range Achievement Test), Gray Oral Reading Tests

Other assessment
Nil

Lesion location
• Case 1: Resection carried across the occipital lobe from medial to lateral in a coronal plane (4.5 cm forward along the surface from the occipital tip)
• Case 2: Occipital lobectomy begun 6 cm from the occipital pole and carried into the occipital horn of the lateral ventricle. Bleeding led to clipping of a posterior medial branch of the posterior cerebral artery. Partial dorsal occipital lobectomy was completed

Lesion type
Cases 1 and 2: Lobectomy for the resection of tumors

Language
English

Lexical access via letter naming in a profoundly alexia and anomic patient: a treatment study

M. L. Greenwald and L. J. Gonzalez-Rothi

Abstract
We report the results of a letter naming treatment designed to facilitate letter-by-letter reading in an aphasic patient with no reading ability. Patient MR’s anoma for written letters reflected two loci of impairment within visual naming: impaired letter activation from print (a deficit commonly seen in pure alexic patients who read letter by letter) and impaired access to phonology via semantics (documented in a severe multimodality anoma). Remarkably, MR retained an excellent ability to pronounce orally spelled words, demonstrating that abstract letter identities could be activated normally via spoken letter names, and also that lexical phonological representations were intact when accessed via spoken letter names. MR’s training in oral naming of written letters resulted in significant improvement in her oral naming of trained letters. Importantly, as MR’s letter naming improved, she became able to employ letter-by-letter reading as a compensatory strategy for oral word reading. MR’s success in letter naming and letter-by-letter reading suggests that other patients with a similar pattern of spared and impaired cognitive abilities may benefit from a similar treatment. Moreover, this study highlights the value of testing the pronunciation of orally spelled words in localizing the source of pre-lexical reading impairment and in predicting the functional outcome of treatment for impaired letter activation in reading.

Journal

Neurocase Reference Number:
P747

Primary diagnosis of interest
Pure alexia and anoma

Author’s designation of case
MR

Key theoretical issue
• An anomic patient trained to orally name written letters became able to employ letter-by-letter reading as compensatory strategy for word reading

Key words: pure alexia; single letter report; oral naming; rehabilitation

Scan, EEG and related measures
MRI

Standardized assessment
Western Aphasia Battery, Boston Naming Test, Battery of Adult Reading Function

Other assessment
Nil

Lesion location
• Left hemisphere involving occipital lobe and extending from temporal region to temporal–parietal–occipital junction

Lesion type
Haemorrhage

Language
English
Reading speed in pure alexia

J. R. Hanley and J. Kay

Abstract
This study investigated possible causes of differences in reading speed between two alexic patients who read words letter by letter. As both patients appeared to rely on serial left-to-right processing of letters within words, the difference in reading speed did not seem to be related to any differences in the extent to which the patients could recognize letters in words in parallel or ‘ends-in’. Differences in reading speed also seemed to be unrelated to the patients’ ability to identify individual letters since their letter recognition accuracy was very similar. Furthermore, although patient PD was significantly slower at reading words aloud than patient DC, PD was in fact significantly quicker than DC on a test that has previously been used to assess letter recognition skills in letter-by-letter readers. It is therefore concluded that PD reads words more slowly because of an additional impairment at the level of the word form system. The results therefore reinforce the distinction between Type 1 and Type 2 letter-by-letter readers that was first drawn by Patterson and Kay.

Journal
Neuropsychologia 1996; 34: 1165–74

Neurocase Reference Number:
P748

Primary diagnosis of interest
Pure alexia

Author’s designation of case
PD (also described in Hanley et al., Neuropsychologia 1992; 30: 237–56, and in Kay et al., Cognitive Neuropsychology 1991; 8: 249–73)
DC

Key theoretical issue
• Differences in reading latencies are not a function of the speed with which individual patients can process letters. Slower readers may suffer from an impairment at the level of the word form system that reduces the accuracy and speed with which a word can be read

Key words: acquired dyslexia; letter-by-letter reading; letter recognition; pure alexia

Scan, EEG and related measures
Case 1: Nil
Case 2: CT

Standardized assessment
Case 1: Nil
Case 2: WAIS, Wechsler, Graded Naming Test, PALPA

Other assessment
Nil

Lesion location
• Case 1: Region of the posterior thalamus
• Case 2: Left posterior temporal and occipital lobes

Lesion type
Case 1: Haemorrhage due to an arteriovenous malformation
Case 2: Haemorrhage

Language
English

A case of ‘sign blindness’ following left occipital damage in a deaf signer

G. Hickok, E. Klima, M. Kritchevsky and U. Bellugi

Abstract
We report on a right-handed, deaf, life long signer who suffered a left posterior cerebral artery (PCA) stroke. The patient presented with right homonymous hemianopia, alexia and a severe sign comprehension deficit. Her production of sign language was, however, virtually normal. We suggest that her syndrome can be characterized as a case of ‘sign blindness’, a disconnection of the intact right hemisphere visual areas from intact left hemisphere language areas. This case provides strong evidence that the neural systems supporting sign language processing are predominantly in the left hemisphere, but also suggests that there are some differences in the neural organization of signed vs spoken language within the left hemisphere.

Journal
Neuropsychologia 1995; 33: 1597–606

Neurocase Reference Number:
P749

Primary diagnosis of interest
Sign blindness, pure alexia

Author’s designation of case
Case LHD-111

Key theoretical issue
• Sign blindness is due to a disconnection between visual information in the intact right visual cortex and the intact language areas in the left hemisphere

Key words: sign blindness; pure alexia; disconnection

Scan, EEG and related measures
CT

Standardized assessment
Salk Sign Diagnostic Aphasia Examination (adapted version of the Boston), Cookie Theft Picture, Benton Facial Recognition

Other assessment
Nil

Lesion location
• Territory of the left posterior cerebral artery

Lesion type
Ischemic infarction

Language
English
Music alexia in a patient with mild pure alexia: disturbed visual perception of nonverbal meaningful figures

T. Horikoshi, Y. Asari, A. Watanabe, Y. Nagaseki, H. Nukui, H. Sasaki and K. Komiya

Abstract
A 26-year-old female pianist suffered from an intracerebral haematoma caused by an arteriovenous malformation of the left occipital parasplenial region, which was operated on 7 months after the onset. Incomplete right hemianopsia, mild pure alexia, and partially disturbed naming of visual objects persisted several months after the removal of the malformation. Evaluation of musical ability 1 and 3 months after surgery showed that her auditory recognition of music was intact. She could sing and play melodies already learned and could dictate well the notes after hearing tones. However, she had difficulty in reading music, especially the pitch of notes, even for simple sequences of four notes. In contrast, her rhythm reading was fairly good. Her visual recognition of other symbolic figures like road signs was also markedly impaired. These results suggest that her visual recognition of written music as well as of other symbolic figures underwent a preliminary verbal decoding in the left hemisphere and that pitch reading was more dependent on verbal processing than rhythm reading.

Journal
Cortex 1997; 33: 187–94

Neurocase Reference Number:
P750

Primary diagnosis of interest
Pure alexia, music alexia

Author’s designation of case
Nil

Key theoretical issue
- Reading music, and, in particular, the pitch of notes, is dependent on verbal processing

Key words: pure alexia; music alexia

Scan, EEG and related measures
CT, SPECT, cerebral angiography, MRI

Standardized assessment
Japanese WAIS-R, Kohs cube test, Standard Language Test for Aphasia of the Japanese Society of Aphasiology

Other assessment
Nil

Lesion location
- Occipito-temporal deep white matter surrounding the trigone, posterior horn of the left lateral ventricle and the splenium of the corpus callosum

Lesion type
Haemorrhage plus hydrocephalus due to arteriovenous malformation

Language
English

Pure alexia and word-meaning deafness in a patient with multiple sclerosis

M. Jonsdottir, T. Magnusson and O. Kjartansson

Abstract
Objective: To describe pure alexia and auditory comprehension problems in a young woman with multiple sclerosis (MS). Patient: A 33-year-old woman with MS who complained of difficulties in reading and comprehending spoken language was referred for a neuropsychological examination. Reading difficulties were confirmed and most of the reading errors were additions, omissions, and substitutions of single letters. While the patient reported that the letters seemed to disappear before her eyes, no general problems with visual attention, visual discrimination, or scanning were detected. No difficulties with spelling were reported. The auditory comprehension deficit is interpreted as a form of a semantic access disorder and is not due to generalized slowing in information processing or conceptual disintegration. Conclusions: Pure alexia is unusual in MS and to our knowledge only 1 other case has been reported (in Japanese). Memory impairments and slowed information processing are probably the most frequent cognitive sequelae of the disease and, consequently, the literature is biased toward the study of those cognitive domains. However, given the wide distribution of sclerotic plaques in MS, it could be argued that we should expect some variability of cognitive changes in MS. Striking deficits as seen in this patient should make us more sensitive to this possibility.

Journal
Archives of Neurology 1998; 55: 1473–4

Neurocase Reference Number:
P751

Primary diagnosis of interest
Pure alexia, word-meaning deafness, multiple sclerosis

Author’s designation of case
Nil

Key theoretical issue
- Because of the wide distribution of sclerotic plaques in multiple sclerosis, cognitive deficits might also include pure alexia and word-meaning deafness

Key words: pure alexia; word-meaning deafness; multiple sclerosis

Scan, EEG and related measures
MRI

Standardized assessment
Token Test

Other assessment
Nil

Lesion location
- Bilateral periventricular white matter, bilateral lesions in occipital lobe white matter and small bilateral subcortical lesions in superior temporal gyri

Lesion type
Infarction

Language
English
Pure alexia in a Kannada–English bilingual

P. Karanth

Abstract
The case history of a 57-year-old Kannada–English bilingual who suddenly lost his ability to read is presented. The relative severity of the reading disorder at various levels, in Kannada and English, as well as its recovery is described. The possible reasons for the findings as well as their implications for the interpretation of alexia as a total inability to read are discussed.

Journal

Neurocase Reference Number:
P752

Primary diagnosis of interest
Pure alexia

Author’s designation of case
NR

Key theoretical issue

- The patient was more impaired in reading Kannada than English, although Kannada was his first written language. This may be due to the fact that the orthography of Kannada is much more complicated than that of English, with 14 possible graphemes for every consonant.

Key words: pure alexia; bilingual reading

Scan, EEG and related measures

- No mention

Standardized assessment
Nil

Other assessment
Nil

Lesion location

- No mention

Lesion type
No mention

Language
English

Cross-modality word matching in letter-by-letter readers

R. B. Katz

Abstract
Letter-by-letter readers can match spoken words to printed words they cannot read. Good word-matching ability may be attributable to a priming effect. However, since letter-by-letter readers have intact spelling ability, an alternative hypothesis is that word matching depends on a strategy of scanning printed words for recognizable letters known to be in the target items. In the present study, experimental manipulations that taxed the ability of the two subjects to process key letters (those that distinguished target words from foils) or scan for them caused a decline in word-matching performance. Thus, letter-by-letter readers may rely on sequential letter processing to accomplish cross-modality word matching.

Journal
Cortex 1990; 26: 65–76

Neurocase Reference Number:
P753

Primary diagnosis of interest
Letter-by-letter reading

Author’s designation of case
Case 1: RH
Case 2: JW

Key theoretical issue

- Pure alexic patients rely on sequential letter processing to match a spoken word to its printed counterpart. There is no cross-modal priming effect per se.

Key words: letter-by-letter reading; cross-modality priming; sequential letter processing

Scan, EEG and related measures
Cases 1 and 2: CT

Standardized assessment

Case 1: Boston Diagnostic Aphasia Examination, Boston Naming Test, Wechsler, Wide Range Achievement Test (WRAT), Peabody Picture Vocabulary Test
Case 2: Boston Diagnostic Aphasia Examination, Boston Naming Test, WRAT

Other assessment
Nil

Lesion location

- Case 1: Old infarct in the left fronto-temporal region, two watershed cerebellar infarcts, and an acute left occipital involving the region of the posterior cerebral artery
- Case 2: Left occipital lobe, the two cerebellar hemispheres, left temporal lobe extending to the thalamus

Lesion type
Case 1: CVA suspected
Case 2: CVA

Language
English
Simultaneous form perception in a case of letter-by-letter reading

J. Kay and R. Hanley

Abstract
The paper examines the performance of a letter-by-letter reader, PD, on tests which involve the perception of words and letter-strings under tachistoscopic presentation conditions. Unlike the patients recently reported by Bub et al. (Brain and Language 1989; 36: 357–76), and Reuter-Lorenz and Brunn (Cognitive Neuropsychology 1990; 7: 1–20), PD shows no evidence of processing the letters of a word in an 'ends-in' fashion. Instead, letters appear to be recognized serially from left to right. Furthermore, PD does not show an advantage of words over pseudowords when asked to report the letters from brieﬂy presented letter strings. In addition, unlike the control subjects, he does not show a word superiority effect in same/different judgements about two simultaneously presented letter-strings (Friedrich et al., Cognitive Neuropsychology 1985; 2: 253–64). A final experiment reveals that his ability to recognize letters under sequential processing conditions is relatively unimpaired, as is his ability to match letters on the basis of physical features with simultaneous presentation. It is suggested that the precise locus of the deﬁcit which precipitates letter-by-letter reading differs from one case to another, and that PD’s primary impairment is caused by an inability to identify more than one letter at the same time.

Journal
Cognitive Neuropsychology 1991; 8: 249–73

Neurocase Reference Number:
P754

Primary diagnosis of interest
Letter-by-letter reading

Author’s designation of case

Key theoretical issue
Letter-by-letter reading is a strategy that is used to compensate for subtly different types of reading impairment in individual cases. PD seems to suffer from the inability to identify letters in parallel

Key words: letter-by-letter reading; serial letter recognition; word superiority effect

Scan, EEG and related measures
CT

Standardized assessment
PALPA, Warrington Recognition Memory Test for Faces, Bradley’s Test for Phonological Awareness, Graded Naming Test

Other assessment
Nil

Lesion location
Region of the left posterior thalamus

Lesion type
Infarction following an haemorrhage due to arterio-venous malformation

Language
English

The localizing significance of limited simultaneous form perception

M. Kinsbourne and E. K. Warrington

Abstract
A patient presenting with an isolated ‘spelling dyslexia’ and impairment in picture interpretation (simultanagnosia) was shown by tachistoscopic studies to have a pathological limitation of simultaneous form perception. At autopsy, a localized lesion was found within the inferior part of left occipital lobe. These findings are discussed in relation to the problem of the disorder of function underlying ‘agnosic alexia’.

Journal
Brain 1963; 86: 697–705

Neurocase Reference Number:
P755

Primary diagnosis of interest
Spelling dyslexia, simultanagnosia

Author’s designation of case
Mrs. B (N.H. 98784)

Key theoretical issue
Pure alexia may be due to a limitation of simultaneous form perception that is not confined to the perception of words, but extends to the perception of any visual array

Key words: pure alexia; simultanagnosia; perceptual deﬁcit

Scan, EEG and related measures
EEG, lumbar air encephalogram, autopsy

Standardized assessment
Babcock sentence, Wechsler, Weigl’s sorting test

Other assessment
Nil

Lesion location
First lesion located 1 cm deep and extended 1 cm anteroposteriorly in the superior temporal gyrus. Second lesion extended backward, from the inferior temporal gyrus to within 1 cm of the occipital pole. This lesion involved the area of brain deep to its superficial marking to a depth of 2.5 cm

Lesion type
Haemorrhage

Language
English
Recovery from alexia without agraphia: report of an autopsy

M. Kurachi, N. Yamaguchi, T. Inasaka and H. Torii

Abstract
The patient is a 58-year-old Japanese teacher of German literature who suffered twice from cerebrovascular accidents, showing alexia without agraphia. Pathological examination showed an old infarct in the posterior two-thirds of the fusiform and almost the whole lingual gyrus, involving the posterior border of the parahippocampal gyrus in the left hemisphere. Left cuneus and the calcarine cortex were preserved. There was degeneration of the lower third of the splenium of the corpus callosum, extending to its occipital radiation and tapetum on both sides. Comparing clinico-pathological findings of the 31 known autopsy cases, it was proposed that the lesion of the left spleno-lingual system produces alexia without agraphia but it may ameliorate. In addition, when spleno-cuneate system is also involved alexia becomes persistent and it may accompany object agnosia or optic aphasia.

Journal
Cortex 1979; 15: 297–312

Neurocase Reference Number: P756

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
KTA

Key theoretical issue
A pure alexic patient with a lesion of the left spleno-lingual system can recover reading. However, when the spleno-cuneate system is also involved, pure alexia becomes persistent.

Key words: pure alexia; autopsy; recovery

Scan, EEG and related measures
EEG, autopsy

Standardized assessment
Wechsler, Hand Eye Ear Test (Head), Poppelreuter’s Hidden Figure Test

Other assessment
Nil

Lesion location
Posterior two-thirds of the fusiform and almost the whole lingual gyrus, involving the posterior border of the parahippocampal gyrus in the left hemisphere. Degeneration of the lower third of the splenium of the corpus callosum, extending to its occipital radiation and tapetum on both sides

Lesion type
Two CVA

Language
English

A study of the visual defect in verbal alexia-simultanagnosia

D. N. Levine and R. Calvanio

Abstract
Three patients with lesions of the left occipito-temporal lobes and the clinical picture of verbal alexia–simultanagnosia are presented. In tachistoscopic experiments, the patients identified letters at very short, normal exposure durations. Aftercoming ‘masking’ patterns did not impair their ability to name single letters more than for normal subjects. When presented with three letters simultaneously, the patients would name only one or two even with very long exposures. If told in advance which letter to name, they successfully reported any single letter of the trigram except for varying difficulty with the middle letter. If told only after the exposure which letter to name, they performed poorly. If they were required to state only whether the three letters were identical, performance was better than for naming. Errors occurred when the odd letter resembled the other two morphologically. The naming of letters of a trigram was better when the letters formed a word than when they did not. Errors consisted of morphological confusions and transpositions of other elements of the trigram. These experimental results can be interpreted as a deficit in the perceptual analysis of compound visual arrays.

Journal
Brain 1978; 101: 65–81

Neurocase Reference Number: P757

Primary diagnosis of interest
Verbal alexia, simultanagnosia

Author’s designation of case
CC
RT
GC

Key theoretical issue
These patients have a deficit in visual perception. Perceptual analysis—the read-out of information from iconic storage into the visual recognition buffer—is impaired.

Key words: pure alexia; simultanagnosia; perceptual deficit

Scan, EEG and related measures
Case 1: CT, technetium brain scan, arteriography, EEG
Case 2: CT, EEG
Case 3: CT, arteriography

Standardized assessment
Cases 1, 2 and 3: Gray Oral, Rey–Osterrieth figure

Other assessment
Nil

Lesion location
Case 1: Left occipital lobe extending to infero-medial temporal lobe
Case 2: Left occipito-temporal region
Case 3: Left occipital cortisectomy located 6 cm above the transverse sinus and 2.5 cm left of the midline. The incision was parasagittal of length 4 cm

Lesion type
Case 1: Infarction secondary to the occlusion of the circummesencephalic portion of the left posterior cerebral artery
Case 2: Haematoma
Case 3: Resection of an arteriovenous malformation followed by the removal of necrotic brain tissue and fresh haematoma

Language
English
Transitory alexia without agraphia in an HIV-positive patient suffering from toxoplasma encephalitis: a case report

C. Lüscher and F. F. Horber

Abstract
An HIV-positive patient presented the classical syndrome of pure alexia. Neuroradiologic investigation by computed tomography showed a ring-like lesion in the left posterior white matter. The clinical manifestation as well as the radiologic findings resolved after antiprotozoal treatment.

Journal
European Neurology 1992; 32: 26–7

Neurocase Reference Number:
P758

Primary diagnosis of interest
Pure alexia without agraphia

Author’s designation of case
Nil

Key theoretical issue
- An HIV-positive patient exhibited pure alexia as presenting symptom of toxoplasma encephalitis. Clinical and radiologic recovery followed antiprotozoal treatment

Key words: pure alexia; HIV; Acquired Immunodeficiency Syndrome; cerebral toxoplasmosis

Scan, EEG and related measures
CT

Standardized assessment
Nil

Other assessment
Nil

Lesion location
- Left posterior white matter involving the radiation and splenium of corpus callosum

Lesion type
Cerebral toxoplasma encephalitis

Language
English

Rehabilitation of a case of pure Alexia: exploiting residual abilities

L. M. Maher, M. C. Clayton, A. M. Barrett, D. Schober-Peterson and L. J. Gonzalez-Rothi

Abstract
We present a case study of a 43-year-old woman with chronic and stable pure alexia. Using a multiple baseline design we report the results of two different interventions to improve reading. First, a restitutive treatment approach using an implicit semantic access strategy was attempted. This approach was designed to exploit privileged access to lexical–semantic representations and met with little success. Treatment was then switched to a substitutive treatment strategy, which involved using the patient’s finger to pretend to copy the letters in words and sentences. Using this motor cross-cuing strategy was 100% accurate and doubled in speed after 4 weeks of intervention. We propose that this patient’s inability to benefit from the implicit semantic access treatment approach may be in part related to her inability to suppress the segmental letter identification process of word recognition.

Journal

Neurocase Reference Number:
P759

Primary diagnosis of interest
Pure alexia

Author’s designation of case
VT

Key theoretical issue
- The patient did not benefit from therapy designed to exploit privileged, covert activation of lexical information but did benefit from a motor cross-cuing strategy

Key words: pure alexia; rehabilitation

Scan, EEG and related measures
MRI

Standardized assessment
Western Aphasia Battery, Boston Naming Test, Boston Diagnostic Aphasia Examination, Battery of Adult Reading Function, Visual Object and Space Perception Battery, Motor-Free Visual Perception Test–Vertical, Psycholinguistic Assessment of Language Processing in Aphasia

Other assessment
Nil

Lesion location
- Left occipital lobe, including lingual gyrus, parts of fusiform gyrus and cuneus, and retrosplenial area

Lesion type
Infarction

Language
English
Anatomical and clinical correlations in a case of alexia without agraphia

F. Michel, B. Schott, M. Boucher and N. Kopp

Abstract
The authors report the anatomical and clinical findings observed in a case of alexia without agraphia due to left posterior cerebral artery infarction, in which there were lesions of the occipital and thalamic regions, the splenium of the corpus callosum, and the hippocampus. They discuss, more particularly, the memory difficulties, and the objects naming trouble.

Journal

Neurocase Reference Number: P760

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
Monsieur P.

Key theoretical issue
- The authors try to associate the patient’s behaviour with their anatomical observations

Key words: alexia without agraphia; memory deficit; visual agnosia

Scan, EEG and related measures
Autopsy

Standardized assessment
Nil

Other assessment
Nil

Lesion location
- In the left hemisphere: splenium of the corpus callosum, forceps major, forceps minor, fusiform and lingual gyri, calcarine circonvolution, hippocampus, inferior longitudinal tract, and optic radiations

Lesion type
Infarction

Language
French

Varieties of pure alexia: the case of failure to access graphemic representations

M. Miozzo and A. Caramazza

Abstract
We document the case of a patient (GV) who, following a left posterior brain lesion, showed a selective and severe deficit in naming visual objects and in reading letters, words, and numerals. Three sets of findings are critical for the interpretation of the patient’s alexia. First, despite intact visual processing abilities and preserved ability to recognize the shape and orientation of letters, GV could not determine whether a pair of letters had the same name. Second, she should not access the orthographic structure and meaning of visually presented words, although she could access meaning from orally spelled words and she could access orthographic structure from meaning in written words. Third, GV could access partial semantic information from pictures and Arabic numerals. Based on this pattern of results, we conclude that the form of alexia manifested by our patient results from failure to access the graphemic representations of letters and words from normally processed visual input. The findings further suggest that access to letter forms and grapheme representations are sequentially ordered stages of processing in word recognition. The results also suggest that graphemic processing may be a distinct property of the left hemisphere.

Journal

Neurocase Reference Number: P761

Primary diagnosis of interest
Letter-by-letter reading, pure alexia, visual agnosia

Author’s designation of case
GV

Key theoretical issue
- Alexia results from a failure to access the graphemic representations of letters and words

Key words: letter-by-letter reading; pure alexia; letter deficit

Scan, EEG and related measures
CT

Standardized assessment
Nil

Other assessment
Nil

Lesion location
- Left occipital and posterior temporal areas extending to the corpus callosum, hypodensity in the right parietal region

Lesion type
Infarction

Language
English
Pure alexia in Japanese and agraphia without alexia in Kanji

H. Mochizuki and R. Ohtomo

Abstract
A 60-year-old right-handed Japanese man with an infarction of the left occipital lobe and inferior temporal gyrus initially showed pure alexia in Kana and Kanji. Later, though pure alexia in Kana persisted, his Kanji reading improved markedly, but with little improvement of Kanji writing. We speculate that different pathways are involved in Kanji reading and writing. Wernicke’s area and its surrounding left middle temporal lobe might play the most important role for Kanji reading when visual information is transmitted by any pathway. The pathway from Wernicke’s area to the left occipital lobe via the middle and inferior temporal pathways may be indispensable for Kanji writing. We postulate ‘agraphia without alexia in Kanji’ due to left inferior subcortical damage.

Journal
Archives of Neurology 1988; 45: 1157–9

Neurocase Reference Number:
P762

Primary diagnosis of interest
Pure alexia, agraphia without alexia

Author’s designation of case
Nil

Key theoretical issue
● Different pathways may underlie Kanji reading and writing with Wernicke’s area and surrounding temporal lobe contributing to reading but pathways from Wernicke’s area to the left occipital lobe via middle and inferior temporal pathways may be important for writing

Key words: pure alexia; Japanese; agraphia without alexia

Scan, EEG and related measures
CT, MRI

Standardized assessment
Nil

Other assessment
Nil

Lesion location
● Left occipital and subcortical structures and inferior temporal gyrus

Lesion type
Infarction

Language
English

Phonological activation in pure alexia

M. Montant and M. Behrmann

Abstract
Pure alexia is a reading impairment in which patients appear to read letter-by-letter. This disorder is typically accounted for in terms of a peripheral deficit that occurs early on in the reading system, prior to the activation of orthographic word representations. The peripheral interpretation of pure alexia has recently been challenged by the phonological deficit hypothesis which claims that a post-lexical disconnection between orthographic and phonological information is responsible for the disorder. Because this hypothesis was mainly supported by data from a single patient (IH), who also has surface dyslexia, the present study re-examined this hypothesis with another pure alexic patient (EL). In contrast to patient IH, EL did not show any evidence of a phonological deficit. Her pattern of performance in naming was not qualitatively different from that of normal readers; she appeared to be reading via a mode of processing resulting in strong serial effects (e.g. onset effect) and strong lexical effects (e.g. frequency effect), a pattern often observed in normal individuals reading unfamiliar stimuli. The present results do not support the phonological hypothesis and are more consistent with peripheral interpretations of pure alexia. The peripheral and the phonological accounts of pure alexia are discussed in light of two current models of visual word recognition.

Journal
Cognitive Neuropsychology 2000: 17; in press

Neurocase Reference Number:
P763

Primary diagnosis of interest
Pure alexia

Author’s designation of case
EL (same patient as Behrmann et al., Neuropsychologia 1998; 36: 1115–32)

Key theoretical issue
● The hypothesis of a phonological deficit as being responsible for pure alexia is tested. EL does not show any evidence of a phonological deficit. Her pattern of reading performance suggests that she is using a slow but normal reading system

Key words: pure alexia; phonological effects; subword processing; serial effects

Scan, EEG and related measures
CT, MRI, angiogram

Standardized assessment

Other assessment
Non-word naming task from Berndt et al. (Cognitive Neuropsychology 1996; 13: 763–801)

Lesion location
● Left occipital and temporal cortices including peristriate inferotemporal association cortex and lateral posterolateral temporal neocortex

Lesion type
Infarction

Language
English
Pure alexia and the viewing position effect

M. Montant, T. Nazir and M. Poncet

Abstract
In the present article, we investigated the reading ability of CP, a pure alexia patient, using an experimental paradigm that is known to elicit the viewing position effect in normal readers. The viewing position effect consists of a systematic variation of word recognition performance as a function of fixation location within a word: word recognition is best when the eyes fixate slightly left from the word centre and decreases when the eyes deviate from this optimal viewing position. A mathematical model (Nazir et al., Bulletin of the Psychonomic Society 1991; 29: 171–4), which provides a good description and quantification of the prototypical shape of the viewing position effect, served to interpret CP’s reading performance. The results showed that, like normal readers, CP was able to process all letters of a word in one fixation. However, in contrast to normal readers, reading performance was optimal when CP was fixating the right half of the word. This somewhat abnormal pattern of performance was due to (1) poor perceptual processing in the right visual field, and (2) poor processing of letters situated towards the end of the word, independent of visual field presentation. A similar pattern of performance was obtained in normal readers under experimental conditions in which lexical knowledge was of restricted use. We suggest that CP’s reading impairment stems from a dysfunction in the coupling between incoming visual information and stored lexical information. This dysfunction is thought to uncover a pre-lexical stage of word processing where letter information is weighted differently as a function of letter position in a word-centred space.

Journal

Neurocase Reference Number: P764

Primary diagnosis of interest
Pure alexia

Author’s designation of case
CP

Key theoretical issue
• The patient’s impairment stems from a dysfunction in the coupling between visual information and lexical information. This dysfunction uncovers a pre-lexical word-centred level of word processing that exists in normal readers

Key words: pure alexia; viewing position effect; word-centred space

Scan, EEG and related measures
CT, MRI

Standardized assessment
Nil

Other assessment
Nil

Lesion location
• Haematoma in the left lingual gyrus, subcortical white matter of the calcarine area

Lesion type
Infarction

Language
English

Can treatment for pure alexia improve letter-by-letter reading speed without sacrificing accuracy?

S. Nitzberg Lott and R. B. Friedman

Abstract
An experimental treatment study designed to improve both the accuracy and the speed of reading was administered to a patient with pure alexia and impaired letter naming. The study focused on the use of letter-by-letter reading. A two-stage approach was employed. The first stage implemented a tactile–kinesthetic strategy to improve accuracy. The second stage concentrated on speed. At the end of the treatment, patient DL was reading both trained and untrained words more accurately and with considerably greater speed than prior to treatment. Accuracy and speed of reading at the sentence level improved as well.

Journal
Brain and Language 1999; 67: 188–201

Neurocase Reference Number: P765

Primary diagnosis of interest
Letter-by-letter reading, pure alexia

Author’s designation of case
DL

Key theoretical issue
• Tactile–kinesthetic re-education can improve letter-by-letter reading

Key words: letter-by-letter reading; tactile–kinesthetic re-education

Scan, EEG and related measures
CT

Standardized assessment
Boston

Other assessment
Nil

Lesion location
• Left posterior temporal occipital lobe infarct, old infarct in the right frontal–parietal lobe, small lacune in the posterior right basal ganglia region

Lesion type
CVA

Language
English
Evolution of a form of pure alexia without agraphia in a child sustaining occipital lobe infarction at 2$^{1/2}$ years

A. E. O’Hare, G. N. Dutton, D. Green and R. Coull

Abstract
The progress of cognitive visual dysfunction over an 8-year period of a child who sustained bilateral occipital lobe infarctions at the age of 2$^{1/2}$ years is described. She survived with normal intelligence and went on to attend mainstream school. She manifested many features of cognitive visual impairment and, in particular, developed a form of pure alexia without agraphia. She achieved some letter-by-letter reading but no sight vocabulary development, including to her own name. She learned to write imaginatively employing phonetically true spelling but cannot read what she has written. Her progress and the difficulties encountered during the management of her condition are discussed in this first case report of the evolution of pure alexia without agraphia in childhood. The features of this syndrome in the developing child who has never developed the capacity to read are contrasted with those seen in affected adults.

Journal

Neurocase Reference Number: P766

Primary diagnosis of interest
Pure alexia without agraphia

Author’s designation of case
Nil (same patient as Dutton et al., Eye 1996; 10: 302–9)

Key theoretical issue
- Despite improved visual acuity after extensive bilateral occipital lobe infarctions, this child exhibited several perceptual deficits as well as pure alexia. Reading was not successful using tactile input

Key words: pure alexia; child; tactile input

Scan, EEG and related measures
CT

Standardized assessment
Vernier Catford Drum, William IQ test, Stycar single optotype assessment, Bust Assessment Cards, Schonell Spelling test, Preferential Acuity card procedure, Ishihara Coloured Vision Plate test

Other assessment
Nil

Lesion location
- Bilateral occipital lobes, more marked on right than left

Lesion type
Infarction

Language
English

Letter dyslexia in a letter-by-letter reader

R. Perri, P. Bartolomeo and M. C. Silveri

Abstract
We describe a letter-by-letter patient who produced misreading errors in both letters in isolation and in words. All errors were visual in nature. We hypothesized an access deficit to the abstract visual representation of letters that prevents letter identification. This deficit could account for the patient’s letter-by-letter behaviour, since each letter constituted a potential identification problem. An access deficit, moreover, could also explain the patient’s letter visual errors. In access processing, in fact, the letters sharing common structural features in their abstract representations were the ones more frequently mismatched.

Journal

Neurocase Reference Number: P767

Primary diagnosis of interest
Letter-by-letter reading

Author’s designation of case
SP

Key theoretical issue
- This patient has a high-level perceptual disorder, that is not specific to orthographic material and that consists in the inability to match the product of visual analysis (letters for example) to the relevant mental images. Difficulties in single letter reading may predict the difficulties in word reading

Key words: letter-by-letter reading; letter deficit; visual errors

Scan, EEG and related measures
CT

Standardized assessment
Nil

Other assessment
Test battery for language examination (Miceli et al., 1991, Milano: Associazione per le ricerche neuropsicologiche)

Lesion location
- Left occipital parietal region

Lesion type
Haemorrhage

Language
English
Primming effects in a letter-by-letter reader depend upon access to the word form system


Abstract
Several types of cognitive and neuropsychological evidence suggest that priming effects on such implicit memory tests as word identification are mediated by a pre-semantic visual word form system that can operate independently of episodic memory. We investigate priming in a letter-by-letter reader, PT, whose pattern of performance on neuropsychological tests indicates that despite PT’s great difficulty in identifying non-studied words. Experiment 2 showed that the priming effect was modality specific whereas Experiment 3 indicated that recall of previously studied words was not modality specific, thus indicating that the observed priming could not be attributed to explicit memory strategies. Experiment 4 revealed no priming of illegal non-words on a letter identification test. The results support the notion that priming on the word identification test depends on access to the word form system.

Journal

Perceptual cues in pure alexia

E. B. Sekuler and M. Behrmann

Abstract
This study provides evidence that pure alexia, or letter-by-letter reading, may be attributed to a general perceptual disorder. The perceptual problem may be unmasked when appropriate perceptual cues are not available to aid in the derivation of an integrated structural description. Four pure alexic patients and eight non-brain-damaged controls participated in this study. In the first two experiments, subjects' reading abilities were assessed on a naming latency and lexical decision task. Experiment 3 replicated Farah and Wallace’s (1991) results that the pure alexia deficit was not specific to orthography. Experiments 4 and 5 further explored the nature of the perceptual disorder using non-orthographic stimuli. In Experiment 4, patient performance on a target detection task was unaffected by the number of parts comprising the object but was unimpaired when the perceptual cue of good continuation was absent. Patient performance also declined when the perceptual cue of symmetry was not available to aid in the integration of occluded object parts in Experiment 5. Overall, the results imply that pure alexia is most likely to arise from a more general, non-orthographic deficit, and that the nature of the disorder is revealed when the perceptual context lacks strong perceptual cues.

Journal
Cognitive Neuropsychology 1996; 13: 941–74

Case 1: Pure alexia

Author’s designation of case
PT

Key theoretical issue
- The word form system is spared in the pure alexic patients who show evidence of priming

Key words: letter-by-letter reading; priming; word form system

Scan, EEG and related measures
CT

Standardized assessment
Western Aphasia Battery

Other assessment
Nil

Lesion location
- Left temporo-occipital region

Lesion type
Infarction

Language
English
Lesion location
• Case 1: No focal lesion; bilateral frontal slowing on EEG. Mildly decreased cerebral perfusion bilaterally on SPECT
• Case 2: Left occipital and temporal lobe
• Case 3: Left occipital lobe
• Case 4: Left occipital lobe

Lesion type
Case 1: Closed head injury
Case 2: Resected arteriovenous malformation
Cases 3 and 4: Infarction

Language
English

Spelling dyslexia: a form of cross-cuing

L. J. Speedie, L. J. Rothi and K. M. Heilman

Abstract
A patient with spelling dyslexia maintained the ability to write, spell, and pronounce spelled nonsense words and words that require knowledge of orthographic rules of language. He was unable to perform grapheme–phoneme conversions except after naming the letter involved. We propose that he uses a letter-naming strategy to circumvent the disconnection of visual areas from the area of visual word images and that his letter-naming strategy represents a compensatory cross-cuing strategy.

Journal
Brain and Language 1982; 15: 340-52

Neurocase Reference Number:
P770

Primary diagnosis of interest
Spelling dyslexia

Author’s designation of case
None

Key theoretical issue
• Visual input of graphic information was isolated from an intact area of visual word images containing the morphophonological rules of spoken language. Verbalization of letters substitutes for visual input

Key words: spelling dyslexia; compensatory strategy

Scan, EEG and related measures
CT, EEG

Standardized assessment
Boston Naming Test

Other assessment
Nil

Lesion location
• Occipital region

Lesion type
Infarction

Language
English
Functional disconnection in pure alexia and colour naming deficit demonstrated by facilitation methods

F.-J. Stachowiak and K. Poeck

Abstract
The authors suggest that the possible combinations of pure alexia with concomitant neuropsychological symptoms form a continuum determined by the extent and severity of the brain lesion. The disconnection mechanisms underlying these syndromes are demonstrated by the facilitating effects of unblocking methods (in the tactile, somesthesic, auditory, and visual modality) on the reading performance of a pertinent case. In the unblocking situation, other pathways than the one impaired by the brain lesion are used. It is proposed that the colour naming deficit is associated with defective verbal identification of graphemic stimuli because of their common linguistic features. The delineation from aphasic colour naming and reading deficit is discussed.

Journal
Brain and Language 1976; 3: 135–43

Neurocase Reference Number:
P771

Primary diagnosis of interest
Pure alexia

Author’s designation of case
Nil

Key theoretical issue
• Pure alexia is due to a disconnection between the intact right visual cortex and the left language cortex. Visual word recognition can be facilitated by using acoustic, tactile or somesthesic stimuli

Key words: pure alexia; colour naming; disconnection; facilitation

Scan, EEG and related measures
EEG, carotid angiography

Standardized assessment
Wechsler, Token Test, Benton

Other assessment
Nil

Lesion location
• Nil

Lesion type
CVA

Language
English

A critical appraisal of neuropsychological correlates of Japanese ideogram (Kanji) and phonogram (Kana) reading

M. Sugishita, K. Otomo, S. Kabe and K. Yunoki

Abstract
Owing to the Japanese language's unique writing system, which consists of phonograms and ideograms, reading impairments of Japanese brain-damaged patients have attracted the interest of many researchers. Past case reports as well as some widely accepted handbooks and textbooks have concluded that a specific aphasia type or lesion site is associated with a particular impairment pattern of phonograms and ideograms in reading. However, the methodology and analytical procedures in previous studies were inadequate for reliable generalizations to be made. First, the test materials were unspecified or inappropriate, or the number of test items was small. Secondly, the conclusions were presented without providing individual performance data to support them. Thirdly, in associating patterns of reading impairment with lesion sites, only single cases were reported. The present investigation was designed to overcome the omissions of previous studies, and examined the ability to read 46 single phonograms and 46 single ideograms aloud in four groups of sufficiently large numbers of patients; namely, seven pure alexics, 23 Broca aphasics, 13 Wernicke aphasics, and seven patients with alexia and agraphia. Ours are the first data to demonstrate unequivocally no consistent linkage between aphasia type and the patterns of impairment of phonogram and ideogram reading. The impairment patterns were not uniform across patients even in the same aphasia group. A majority of the cases in each group showed that phonograms and ideograms were unselectively impaired. However, ideogram reading was more difficult in three cases in the pure alexia and Broca aphasia groups, respectively, and in one case in the Wernicke aphasia group. Phonogram reading was more severely disturbed in four cases among the Broca aphasics and in one case among the patients with alexia with agraphia. An apparent variability of impairment patterns characterized the Broca aphasic group. These dyslexic patterns did not appear to correlate with the site and extent of lesions identified by computerized tomography scans. Past reports linking a particular impairment pattern of phonogram and ideogram reading and a specific lesion site were studies of single cases, and their conclusions seem oversimplified. While sensory and motor dysfunctions can usually be neuroanatomically localized in individuals, impairments of certain high cortical functions, such as the reading of phonograms and ideograms, may not be correlated with damage to definite neuroanatomical structures.

Journal
Brain 1992; 115: 1563–85

Neurocase Reference Number:
P772

Primary diagnosis of interest
Pure alexia, Broca aphasia, Wernicke aphasia, alexia with agraphia

Author’s designation of case
Cases 1–7 (also described: 23 Broca aphasics patients, 13 Wernicke aphasic patients, and seven alexic and agraphic patients)

Key theoretical issue
• Data from 43 patients (among which were seven pure alexic patients) show that the selective impairment of phonogram or ideogram reading is not correlated with damage to a definite neuroanatomical structure

Key words: alexia; aphasia; agraphia; phonograms; ideograms; lesion site

Scan, EEG and related measures
Cases 1–7; CT
Alexia without agraphia, hemianopsia, or colour-naming defect: a disconnection syndrome

F. M. Vincent, C. H. Sadowsky, R. L. Saunders and A. G. Reeves

Abstract
A patient with alexia without agraphia, hemianopsia, or colour-naming defect was found at operation to have a meningioma arising from the tentorium cerebelli that compressed the inferior aspect of the left temporal–occipital junction. It is presumed to have involved only the left ventral visual association cortex and its inferior outflow tracts to the angular gyrus. The input from the right occipital area also was disconnected from the visual language verbal association area by involvement of the ventral outflow of the splenium of the corpus callosum. Preservation of colour naming and matching suggests that these functions are dependent on the integrity of more dorsal occipital association systems.

Journal
Neurology 1977; 27: 689–91

Neurocase Reference Number:
P773

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
Nil

Key theoretical issue
• The ventral occipital association cortex is necessary for reading but it is not essential for naming and matching colours

Key words: alexia without agraphia; disconnection; reading pathway; colour naming

Scan, EEG and related measures
EEG, technetium 99 brain scan, angiography

Standardized assessment
WAIS

Other assessment
Nil

Lesion location
• Inferior portion of the posterior temporal lobe on the left

Lesion type
Mass effect due to a 45 g gross tumor. Extraparenchymal removal of the tumor

Language
English
Effect of print size on reading time in a patient with verbal alexia

B. T. Woods and E. Pöppel

Abstract
The reading performance of a patient with the syndrome of ‘alexia without agraphia’ could be markedly improved by increasing print size and exposure time. In contrast, his recognition of letters, letter groups and object drawings was normal when compared with a control group and independent of size. The data which demonstrate a material specific deficit of ‘visual acuity’ preclude the interpretation of this case of verbal alexia as an agnosia in the ordinary sense of that term.

Journal
Neuropsychologia 1974; 12: 31–41

Neurocase Reference Number: P774

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
AW

Key theoretical issue
Pure alexia is not an agnosic syndrome. The processes responsible for the recognition of different types of visual material are to some degree functionally and even anatomically divergent

Key words: pure alexia; visual agnosia; print size

Scan, EEG and related measures
Arteriogram

Standardized assessment
Oldfield and Wingfield pictures (MRC Psycholinguistics Research unit, Special Report No.PLLU/65/19); Hardy et al.’s screening test for vision (Journal of the Optical Society of America 1946; 36: 610–4); Farnsworth–Munsell 100-Hue Test; Rey–Osterrieth Figure

Other assessment
Nil

Lesion location
Possibly, the territory of the left posterior cerebral artery, involving the medial occipital lobe and either the splenium of the corpus callosum or the fibres emerging from it

Lesion type
Infarction suspected

Language
English

Right unilateral dyscopia of letters in alexia without agraphia

A. Yamadori

Abstract
In two cases of pure alexia with computed tomography (CT) evidence of lesions in the left occipital lobe, the left hand copied letters normally, but the right hand copied defectively. The findings reinforce the disconnection theory of alexia without agraphia.

Journal
Neurology 1980; 30: 991–4

Neurocase Reference Number: P775

Primary diagnosis of interest
Alexia without agraphia

Author’s designation of case
Case 1
Case 2

Key theoretical issue
Two pure alexic patients were impaired in copying letters with the right hand. These results reinforce the theory of a disconnection between the intact right visual cortex and the left angular gyrus as responsible for pure alexia

Key words: pure alexia; copying letters; disconnection theory

Scan, EEG and related measures
Case 1 and 2: CT

Standardized assessment
Nil

Other assessment
Nil

Lesion location
Case 1: Medial portion of the left occipital lobe involving part of the hippocampal gyrus, the fusiform gyrus, the cuneus, the lingual gyrus, and the deep white matter underlying them; the splenium of the corpus callosum may be involved
Case 2: Medial portion of the left occipital lobe involving the hippocampal gyrus, the fusiform gyrus, the cuneus, the lingual gyrus, and the deep white matter underlying them; the splenium of the corpus callosum is also involved

Lesion type
Case 1: Occlusion of the left posterior cerebral artery
Case 2: Nil (but the lesion corresponds to the territory of the left posterior cerebral artery)

Language
English