

# Cognitive Primer

## Alexia

### Introduction

The term alexia denotes the presence of an acquired reading disorder that prevents comprehension or production of written language<sup>1</sup>. Although the word “dyslexia” is equivalent, it is often used to refer to a range of disorders seen in people who fail to develop normal reading skills in childhood.

Normal people can read single words as quickly as single letters. Thus, the letters of a word are perceived in parallel, and no eye movements are made within a word unless it is long or unfamiliar. Reading text requires the reader, in addition, to direct their eyes along the array of words. Therefore, during fixation on a word embedded in text, two processes occur in parallel: word recognition, and the planning of the eye movement onto the viewing point of the next word. Specific regions throughout both cerebral hemispheres are involved in supporting the interdependent processes required to achieve effortless text reading (high visual acuity, letter and word recognition, central language processing, directed visuo-spatial attention and oculomotor control)<sup>2</sup>. Any disease process that affects oculomotor control will be likely to impinge on reading ability, but these disorders are beyond the scope of this article; I will instead concentrate on the syndromes caused by disruption to visual processing of word forms or of those neural regions that support central language processing.

The high visual acuity required to discern word forms is in part due to the preferential representation of foveal and parafoveal vision (having radii of 1° and 5° around fixation, respectively) in the primary visual cortex. Bilateral extra-striate cortices are also involved in reading, but the first cortical region to exhibit asymmetric activity to word forms is left ventro-lateral occipito-temporal cortex (VLOT – see figure). This region, along with an area of cortex that extends medially and anteriorly, is often referred to as the ‘visual word-form area’ as damage to it, or its connections, can lead to the isolated alexic syndrome first recorded by Jules Dejerine in 1892 – ‘alexia without agraphia’ also known as ‘pure’ alexia<sup>3,4</sup>. This syndrome is characterised by the breakdown of the patient’s ability to process word forms as a whole unit. As letter identification is often spared, patients tend to read using a letter-by-letter strategy which is laborious and error prone, especially for longer words. There is still debate as to whether this region solely or even predominantly supports reading function<sup>5</sup>, but a patient with pure alexia who writes at a normal pace and then fails to read back to you what they have just written is one of the more striking signs in clinical neurology.

### Classification

Neurologists and psychologists differ in the terminology they use to describe alexic syndromes<sup>6,7</sup>; however, both recognise the importance of identifying whether the syndrome occurs as part of a generalised language disorder (alexia in the context of aphasia; alexia with agraphia; ‘central’ alexia) or in an isolated form (alexia without agraphia; ‘peripheral’ alexia). The neuropsychological classification of central alexia is based around the relative impairment of two functionally distinct reading routes: reading by sound and reading by sight. Normal readers can pronounce unfamiliar words by applying the common rules of correspondence between spelling and sound in their language. These rules work for the majority of English words, but there is a sizable minority of irregular words (e.g. yacht, blood, two and mauve) which cannot

be read in this way and rely on the reader having built up a “sight” vocabulary, whereby words can be read without recourse to “sounding out”. Patients who can only read by the sight route typically have only minor reading deficits, but are unable to read non-words such as “mune” as these words will not be present in their ‘sight’ vocabulary: so-called ‘phonological dyslexics’<sup>8</sup>. Patients who can only read by sound can manage non-words but have problems with irregular words, pronouncing pint as if to rhyme with mint: so-called ‘surface’ dyslexics<sup>9</sup>. Regularisation errors are often seen early in patients presenting with a temporal variant of fronto-temporal dementia (FTD), allowing this condition to be differentiated from both frontal forms of FTD and Alzheimer’s disease<sup>10</sup>. Patients are also described who have a double deficit, affecting, to different degrees, both the sight and sound reading routes; such patients are deemed to have ‘deep’ dyslexia<sup>9,11</sup>. As well as producing error patterns consistent with the two conditions just mentioned, these patients have additional problems reading function words (it, and, when), and often make semantic errors; e.g. a patient with this syndrome read “inn” as “pub”, but was unable to read the word “in” at all (Prof. Richard Wise – personal communication). Patients with central alexia often have their reading disorder ignored by clinicians as comprehension and speech output impairments tend to overshadow any disability caused by the alexia; however, occasionally the aphasic and alexic components dissociate to some degree resulting in grossly aphasic patients being able to read quite well, thus allowing communication to continue through this modality<sup>12</sup>.

In contrast, patients with a peripheral alexia have preserved central language function (although specific anomic deficits have been noted, the commonest being colour anomia, an impairment of naming in the context of normal colour perception). Patients with pure alexia may initially present with global alexia, being unable to recognise and name letters<sup>13</sup>. In hemianopic alexia, word recognition is intact, but the presence of a right homonymous hemianopia leads to disruption of the visuomotor co-ordination of eye movements during text reading<sup>7</sup>. Patients with neglect alexia consistently make errors on either the initial or final letters of a word (e.g. clock read as block)<sup>14</sup>. Another form of peripheral alexia is attentional alexia, where patients complain of letter crowding, sometimes blending elements of two words into one; they perform better when word stimuli are presented in isolation rather than flanked by other words and letters<sup>15</sup>.

When central alexic disorders are due to focal pathology such as stroke, they are almost always associated with damage to the dominant hemisphere in the territory of the middle cerebral artery (MCA). If peripheral alexia is due to focal pathology, then this is usually in the territory of the posterior cerebral artery (PCA) or in the borderzone between MCA and PCA, potentially causing additional ‘parietal’ (perceptual) or ‘temporal’ (associative) agnosias as well as other neuropsychological deficits associated with damage to or disconnection of occipital cortical regions.

### Clinical examination

The general neurological examination may provide clues as to the lesion site and thus the type of alexia that may be expected. If a patient is aphasic then it is important the examiner gets a feel for their speech output, evaluating spontaneous speech (open questions), automatic speech (counting, days of the week) and repetition (single words

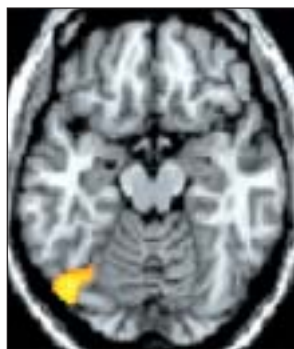


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# Cognitive Primer

Alexia sub-type	Error patterns (specific test material)	Usual lesion site
<b>Central:</b>	<b>Alexia part of a general language disorder</b>	
Reads by sound (surface dyslexia)	Regularisation errors, can read non-words (Irregular words, non-words).	Left medial +/- lateral temporal lobe (FTD).
Reads by sight (phonological dyslexia)	Difficulties with suffixes, unable to read non-words (non-words).	Left temporo-parietal lobe.
Both routes damaged (deep dyslexia)	As above plus: semantic errors, frequency/ imageability effects, poor with function words.	Extensive left temporo-parietal lobe.
<b>Peripheral:</b>	<b>Other language functions usually spared</b>	
Global	Very slow or inaccurate letter naming but can recognise letters (number naming may be spared).	Left VLOT or connections and splenium of CC <sup>13</sup> .
Pure	Slow and inaccurate reading, short words easier, may use a "letter-by-letter" strategy.	Left VLOT or connections.
Hemianopic	Slow but accurate text reading, may miss prefix/suffix depending on side of hemianopia, left HH usually more disabling than right HH (fields).	Any cause of a HH, often macular-splitting, often post-geniculate.
Neglect	Errors on prefix more common than suffix.	Right parietal lobe.
Attentional	Merged words, letter crowding (better with isolated words).	Left parietal lobe.

The main sub-types of alexia are tabulated along with their commonly associated error patterns and lesion sites. Recommended specific test material outside that found in ordinary printed material is bracketed. CC = corpus callosum; HH = homonymous hemianopia; VLOT = Ventro-lateral occipito-temporal cortex



Axial and coronal views of left VLOT cortex activated during an H<sub>2</sub>O<sub>15</sub> PET study. Subjects viewed single words at increasing rates. The group PET data are displayed on a canonical single-subject T1 MRI image<sup>13</sup>. Damage to or disconnection of this region usually causes pure alexia.

and sentences), before testing reading as the main aim in this scenario is to look for relative differences in language function between aural and graphic probes. If an isolated reading disorder is identified, then writing should also be tested (spontaneous and to dictation).

Although a variety of psychological test batteries are available for detailed assessment of reading, (e.g. the PALPA<sup>16</sup>) most clinicians can get a feel for the broad types of alexia with just a pen and paper and some non-specialist reading material (a newspaper or magazine is often to hand and tends to contain different font sizes, simple sentences and pictures; the latter can be used to screen for anomia, hemi-neglect, simultanagnosia, and colour anomia).

Visual acuity and fields need to be checked first. If a hemianopia is present with > 5 degrees of vision preserved to the right of fixation then it is unlikely that there

will be a hemianopic contribution to the reading deficit. Patients should read a simple passage of print out loud with the examiner recording their errors. Errors are usually categorised along two principle axes according to the type of speech error made and the class of word (in terms of grammar and orthography) that the error is made on. Errors of speech output usually fall into two groups: semantic or phonological e.g. "cat" misread as "dog" or "cab" respectively. Phonological errors are responses where the phoneme (speech sound) structure differs from the target enough for a word to have either its meaning changed or lost, and are usually due to phonemic substitutions, insertions or omissions which can occur at the beginning, middle or end of a word. Some patients will tend to make errors on certain grammatical classes of words (nouns, verbs, modifiers or function words), while others will have problems along other descriptive axes

# Cognitive Primer

such as the commonness (frequency), concreteness (imageability), or letter or syllable length of a word.

## Treatment

There are no generally agreed specific therapies for either the central or peripheral alexias, although there is a plethora of case reports detailing successful, uncontrolled, bespoke behavioural interventions. In hemianopic alexia, where the neural representations of written word forms have been spared, the emphasis has been on retraining the oculomotor system in order to improve reading scanpaths<sup>17</sup>. There is an ongoing randomised, controlled study aimed at assessing different oculomotor interventions in this condition (please contact the author for details).

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### Victorian Incurables: A History Of The Royal Hospital For Neuro-Disability, Putney Professor G C Cook

This is the history of an institution originally founded as the Royal Hospital for Incurables, and recently renamed for the last time the Royal Hospital for Neuro-Disability. It was founded in 1854 by Andrew Reed, one of the great (although poorly documented) philanthropists of 19th century Britain. The raison d'être of the institution was provision of long-term care for patients who could not be accommodated in general hospitals. However, its early days were plagued by controversies including the site of the definitive hospital, the ultimate choice being West Hill, Putney. The complex voting system and the absence of women in management were other highly contentious matters, all of which are documented here. Amongst enthusiasts for the initiative were Charles Dickens and Florence Nightingale. Many other famous names of the period are also mentioned, including Joseph Merrick better known as 'The Elephant Man'. Following the introduction of the National Health Service, the board managed to retain voluntary status, and now in its 150th year it has become a model for the care of the chronically sick and a pioneer of rehabilitation. Her Majesty the Queen is currently Patron of the hospital, and the Countess of Lichfield its President.

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