942-5
Alexia without Agraphia
Neurovisual Exam

Visual acuity 20/60 or better OU (made error reading Snellen chart)
Fields: Dense right homonymous hemianopia
Pupils: equal, normal reflexes
Full eye movements
Normal discs and fundi OU
Visual Fields

Figure 1. Automated perimetry showing dense right homonymous hemianopia
Color Vision

Ishihara pseudo-isochromatic test of color vision performed without error
Read single and two digit numbers by tracing them out correctly with his finger
Unable to name color correctly (color anomia)
Named red and blue bottle caps correctly
Named green, purple and yellow, orange
Matched two red objects correctly
Visual Tests

Correctly identified 2 out of 3 famous faces

No visual disorientation - simultanagnosia
Neurological Exam

Slight right facial weakness
A mild right hemiparesis with right pronator drift
Mild hyperreflexia on the right with an extensor plantar
No sensory extinction, apraxia, astereognosis, or agraphesthesia
Stroke Presentation

Alexia with agraphia
A dense right homonymous hemianopia
Color anomia
Neuroimaging

A repeat MRI with DWI showed a new left occipital lobe infarct in the distribution of the left P2 division of the posterior cerebral artery (PCA).
Neuroimaging

Series of axial FLAIR scans shows a posterior cerebral artery infarct involving the occipital lobe and splenium of the corpus callosum.
Neuroimaging
Neuroimaging
Neuroimaging
Disconnection Syndrome

Figure 2. Connections involved in naming a seen object and in reading. The visual pattern is transferred from the visual cortex and association areas to the angular gyrus, which arouses the auditory pattern in the Wernicke area. The auditory pattern is transmitted to the Broca’s areas, where the articulatory form is aroused and transferred to the contiguous face area of the motor cortex.

With destruction of the left visual cortex and splenium (or intervening white matter), words perceived in the right visual cortex cannot cross over to the language areas and the patient cannot read.
Criteria for Diagnosis of Pure Alexia

Severe disturbance of reading comprehension
Linguistically correct writing spontaneously and to dictation
Normal oral spelling
Absence of aphasia and dementia
Damasio’s Criteria for Color Anomia

Damage to the left lingual gyrus
Damage to the left hippocampal region (hippocampus plus parahippocampal region)
Presence of right hemianopia
Psychologists have noticed that pure alexics often read slowly, pronouncing or naming each letter aloud, resulting in an increase in reading time for words as a function of their length.

This has been termed “letter-by-letter reading” and the question it raises is: How is letter and word information transmitted across different parts of the callosum from right to left?
The basic idea has been that letter identities are transmitted in parallel through the splenium and in less efficient, possibly serial ways elsewhere.
Reading

Functional properties
- Lexico-semantic features
- Phonology
- Verbal working memory
- Sequence of abstract letter identities
- Font invariant
- Location invariant

Anatomy and connectivity

Language areas

VWF

R-VWF

V1

V2

V4

W-O-R-D

W d o v o l

b r o W

Word

- Increasing receptive fields
- Increasingly abstract features

Vision-Spatial attention

et al.
Reading

Figure 3. Model of the visual word processing system. Letters are first analyzed through a cascade of contralateral areas ranging from V1 to V4, which compute increasingly abstract representations. Eventually, an invariant representation of letter identities is created in the VWFA, located within the left occipito-temporal sulcus. The VWFA then projects to structures involved in phonological or lexico-semantic processing. Spatial attention (here represented by dark and light gray gradients) modulates the operation of all levels of word processing up to the VWFA, with a right-hemispheric predominance. The right-hemispheric region symmetrical to the VWFA (A-VWFA) is thought to subtend residual letter identification in patients with letter-by-letter reading. Letter identities are then transferred to language areas across the corpus callosum.
Dejerine’s Lesion Drawings

Confrontation of Dejerine’s (1892) original lesion drawings (left column) with fMRI activations in control subjects from the present study (right column). Top row: ventral occipito-temporal lesion responsible for pure alexia, and activations by alphabetic stimuli versus chequerboards. Bottom row: left angular lesion responsible for alexia-with-agaphia (dotted), and activations by real words versus consonant strings.

Cerebral Cortex December 2003, V 13 N 12
Dejerine’s Diagram
## Cases in the Literature

### Major behavioral findings in the patients of Dejerine, Greenblatt, Geschwind, and Mohr

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<th>Right hemiacromatopsia</th>
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# Anatomic Basis of Pure Alexia

## Behavioral findings in 16 patients with pure alexia

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* Patient had left hemiachromatopia. Color naming could not be tested.

RHH Right homonymous hemianopia.
RSQ Right superior quadrantanopia.
References


