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# Progressive ventral posterior cortical degeneration presenting as alexia for music and words

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**Article abstract**—Patients with posterior cortical atrophy may have dorsal visual system (occipital-parietal) dysfunction (optic ataxia, visuospatial disorientation, and simultanagnosia), ventral visual system (occipital-temporal) dysfunction (pure alexia, prosopagnosia, visual anomia, and agnosia), or both. We report a professional musician with ventral system dysfunction whose first symptom was alexia for music. Subsequently, she developed pure alexia for words but had preserved sorting of words. These observations suggest that the ventral visual system is important in music and word reading. However, sorting of words may be mediated by the dorsal visual system.

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In 1988, Benson et al.[1] described five patients with a progressive dementia who had alexia, agraphia, visual agnosia, and components of Balint's, Gerstmann's, and transcortical sensory aphasia syndromes. The term posterior cortical atrophy was used to describe this syndrome because of the prominent posterior atrophy seen on imaging studies in some patients.

Since this report, patients have been described that initially have optic ataxia, visuospatial disorientation, and simultanagnosia.[2,3] Other cases have initially had spelling or letter-by-letter alexia (pure alexia) and visual anomia.[4] As a result, Mackenzie Ross et al.[3] and Caselli[5] proposed a division of these posterior cortical atrophy cases into dorsal ("where" visual system) and ventral ("what" visual system) subtypes. Pathology on such cases is generally regarded to be that of Alzheimer's disease.[3,5] We report a professional musician with the ventral subtype of posterior cortical degeneration whose first symptom was an inability to read music. In addition, although she had a spelling alexia, she could sort words by categories.

**Case report.** A 65-year-old woman who is a professional musician was evaluated at the Memory Disorders Clinic at the University of Florida College of Medicine. Five to 6 years prior, she noticed difficulty sight-reading new music. Subsequently, she began to have more frequent lapses in memory while performing at concerts. However, her memory for music when not having to perform before audiences was relatively preserved. She then developed difficulty

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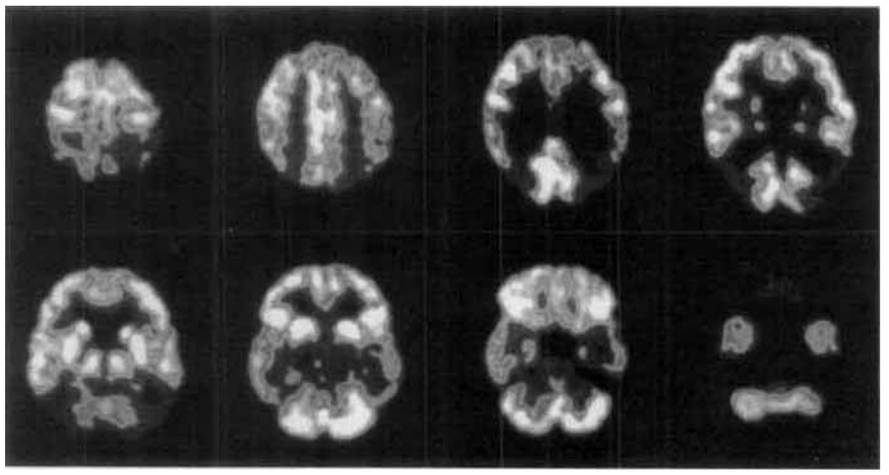

*Figure. 18-Fluorodeoxyglucose PET axial images for this patient. Top left images are superior, bottom right images are inferior (every third image is shown). Images are reversed (left side of brain is on the right side of each image).*

reading text, but writing was preserved. Eventually, she was unable to read her own writing after being distracted. Although formal ophthalmologic evaluation was normal, there were inconsistent reports of a right homonymous field defect. Over the period of 5 to 6 years, her symptoms slowly and steadily progressed to include difficulties with word finding and recent memory. Past medical history was significant for treated hypertension and hypothyroidism. She was also receiving postmenopausal estrogen replacement therapy.

Her general physical examination was unremarkable. Her neurologic examination revealed inconsistent extinction to double simultaneous visual and auditory stimuli (finger rustle) on the right side and bilateral extensor plantar responses without other pyramidal signs.

Although the patient's spontaneous speech was not anomic, she performed the Boston Naming Test[6] poorly, obtaining a score of 20. The patient made primarily visual perceptual (agnosic) errors. The patient also had a mild amnesia (Hopkins Verbal Learning Test[7] Form 1 score of 16) and an impaired ability to copy intersecting pentagons. Although the patient made some mild apraxic errors with her left hand, her finger naming and right-left orientation, comprehension, repetition, and verbal fluency were all normal. Her frontal subcortical functioning was also normal. There was no distractability on visual field testing, no impersistence to eye closure, and no errors on contrasting programs. She had a normal 1-minute word fluency score with the letters FAS (35) and no Parkinson's signs.

Supplementary testing revealed that she had poor recognition of famous faces. Although the patient's writing was normal, her reading was abnormal. She was only able to read the most simple words and used a letter-by-letter strategy. The patient was rapidly presented with cards (approximately one per second) that had words printed on them. The words were presented too quickly to use this letter-by-letter strategy. The patient correctly sorted these words into living or nonliving categories in seven of seven trials. She did not have optic ataxia (an impairment in visually guided movement) or color anomia.

No abnormalities were observed on her MRI, but PET revealed posterior cortical hypometabolism, sparing the calcarine cortex, that was more prominent on the left than on the right (figure).

**Discussion.** Based on studies of nonhuman primates, Ungerleider and Mishkin[8] divided the visual stream into a dorsal "where" and a ventral "what" system. Humans with bilateral dorsal occipital-parietal lesions demonstrate simultanagnosia, optic ataxia, and psychic paralysis of gaze (Balint's syndrome) and elements of Gerstmann's syndrome (agraphia, acalculia, right-left confusion, and finger agnosia) and aphasia. Our patient showed none of these signs. In contrast, patients with ventral occipital-temporal lesions demonstrate alexia without agraphia with letter-by-letter reading, visual object agnosia, and prosopagnosia. Our patient showed all these signs. Based on the preservation of functions mediated by the dorsal visual stream and the malfunction of the functions mediated by the ventral visual stream, our patient appeared to have primarily a ventral posterior cortical degeneration.[3]

Whereas alexia for music has not been reported with degenerative diseases, it has been reported to be associated with alexia for words in patients with strokes, including alexia without agraphia.[9] Since the work of Dejerine, alexia without agraphia has been associated with ventral temporal-occipital lesions. However, using PET, Sergent et al.[10] studied music reading in normal subjects and found activation in the dorsal (occipital-parietal) visual stream. These PET findings appear to be inconsistent with the observations that difficulty with reading music has been associated with alexia without agraphia from stroke and hence ventral visual stream dysfunction.[11] When performing PET studies to localize functions, investigators often use subtraction techniques. The comparison task used by Sergent et al.[10] was having their subjects view dots. Perhaps the visual presentation of dots activated ventral structures sufficiently to obscure the ventral activation induced by reading music. It is possible that reading music requires activation of both the dorsal and ventral visual streams. Whereas written music contains spatial arrays that denote pitch, it also contains symbols that denote elements such as timing, clefs, sharps, and flats. Perhaps the spatial elements are

processed by the dorsal visual stream and the nonspatial elements are processed by the ventral visual stream. Partial support for this postulate comes from the observations of Judd et al.,[11] who reported a composer with pure alexia (alexia without agraphia with letter-by-letter reading) who had trouble reading unfamiliar music but appeared to read pitch better by looking at the relation of each note to its predecessor. Unfortunately, we did not have an opportunity to learn which elements of music reading were defective in our patient, but future studies may be directed at investigating this dissociation.

Coslett and Saffran[12] reported a patient with pure alexia from a left ventral occipital-temporal lesion. They asked their patient to sort words he could not read into edible and nonedible or animal and nonanimal categories. Because this patient with a left hemispheric lesion was able to sort words that he was unable to read, Coslett and Saffran[12] suggested that the ability to sort may be mediated by the patient's preserved right hemisphere. Our patient had behavioral (e.g., prosopagnosia) evidence for bilateral ventral dysfunction. Therefore, it is possible that her preserved sorting may be mediated not by the right hemisphere but rather by the left hemisphere's dorsal visual system. The patient of Coslett and Saffran[12] also had a preserved left hemispheric dorsal visual system. Although functional preservation of the dorsal visual systems suggests that her preserved implicit word processing was mediated dorsally, our patient's PET images were not taken during the sorting procedure and do not help to resolve this issue. Furthermore, one must be cautious in drawing conclusions regarding brain localization from focal cortical degeneration syndromes due to the presence of more widespread dysfunction in these syndromes than in focal lesions such as strokes. Therefore, further studies are needed to test the left dorsal versus the right hemisphere sorting hypotheses.

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