

# Alexia for Braille following bilateral occipital stroke in an early blind woman

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Recent functional imaging and neurophysiologic studies indicate that the occipital cortex may play a role in Braille reading in congenitally and early blind subjects. We report on a woman blind from birth who sustained bilateral occipital damage following an ischemic stroke. Prior to the stroke, the patient was a proficient Braille reader. Following the stroke, she was

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no longer able to read Braille yet her somatosensory perception appeared otherwise to be unchanged. This case supports the emerging evidence for the recruitment of striate and prestriate cortex for Braille reading in early blind subjects. *NeuroReport* 11:237–240 © 2000 Lippincott Williams & Wilkins.

## INTRODUCTION

Recent studies suggest that in early blind subjects, Braille reading is associated with activation of the occipital cortex. In a PET study, Sadato *et al.* [1] found an increase in regional cerebral blood flow (rCBF) in the striate and prestriate cortex in congenitally and early blind subjects during tactile tasks, whereas sighted individuals showed decreased rCBF. Cohen *et al.* [2] showed that when repetitive transcranial magnetic stimulation (rTMS) was used to transiently disrupt the function of the occipital cortex, it interfered with tactile reading of Braille and embossed Roman letters in early blind subjects, but not in sighted control subjects. In the blind subjects, rTMS over striate cortex induced discrimination errors and distorted tactile perception with subjects occasionally reporting feeling 'phantom' Braille dots. This body of evidence demonstrating activation of striate cortex in early blind Braille readers is not, however, unanimous. Büchel *et al.* [3] recently found that congenitally blind subjects show task-specific activation of prestriate visual areas without activation of striate cortex. Clearly further data must be sought to elucidate this apparent incongruity.

Case studies in which blind Braille readers have become alexic following a CNS insult are uncommon, and generally lack precise descriptions of the extent of cortical damage. In the present report, we describe a patient who, following a cerebrovascular accident, became unable to recognize Braille characters. The findings in this patient

further strengthen the notion that the occipital cortex can be recruited to subserve the function of tactile information processing among early blind subjects.

## CASE REPORT

A 63-year-old right-handed woman was blind (with no light perception) since birth due to retrolental fibroplasia. She had been born 6 weeks prematurely, but the preceding pregnancy had been uncomplicated and aside from severe retinal damage, she did not manifest any other long-term effects of premature birth or of newborn intensive care. She had developed normally, achieving developmental milestones at expected ages. She started to learn to read Braille at age 6, and became a proficient reader. She used Braille extensively in college, graduated with a Bachelor's degree, and had been working for the National Radio Station for the Blind in Spain since age 22. At work, she would use Braille to communicate with colleagues, take notes, and write her reports and memos, which amounted to ~4–6 h of Braille use each day. Her reading speed was remarkable, with a rate of 120–150 symbols/min. She used both hands and several fingers in each hand for character recognition.

At age 52, she developed adult onset diabetes, which was controlled to a reasonable degree with diet and hypoglycemic agents, and she had, for several years prior to presentation, required treatment for hypertension. She smoked one pack of cigarettes per day. According to yearly physical examinations she had mild left ventricular hyper-

trophy, thought to be secondary to the hypertension, but experienced no other consequences from her medical problems. Specifically, there was no documentation of diabetic peripheral neuropathy that might have led to difficulties with Braille reading.

One morning, she complained to her co-workers of light-headedness, difficulty swallowing, and motor incoordination. Despite the patient's complaints, her co-workers reported no objective correlates to her sensations. According to the patient, these vague complaints continued for most of the morning, and around noon she reportedly collapsed. Her secretary did not notice any tonic-clonic activity, and initially felt that the patient might have fainted, since there was no cardiorespiratory arrest noted. She was taken to the University Hospital, where she was found to be obtunded, with withdrawal to strong stimuli but no response to verbal orders. Her breathing was shallow, but there was no respiratory compromise. Her pupils were equal and normally reactive to light. No focal signs on the neurological examination were documented in the Emergency Room evaluation. The patient was admitted to the Intensive Care Unit, where, over the course of the following 24 hours she became alert and able to interact normally with her surroundings. The physician noted a normal physical and neurological examination on the second day of her hospitalization. She no longer reported having difficulty swallowing, nor did she feel light-headed. She was able to get up and walk without problems.

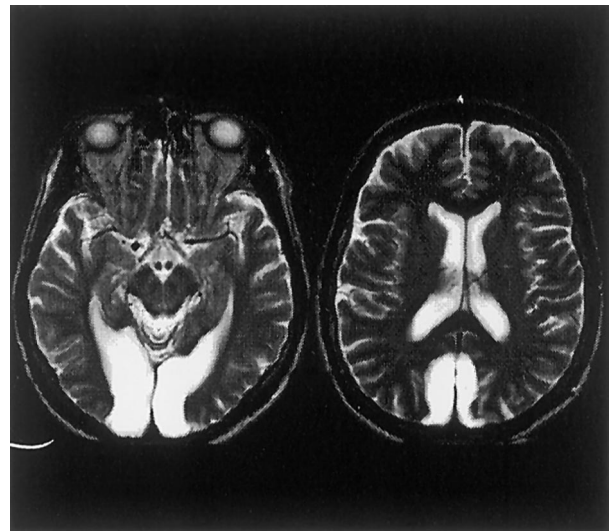
On the second day, when she tried to read a Braille card sent to her, she was unable to do so. She stated that the Braille dots "felt flat" to her, though she described being able to concentrate and determine whether or not there was a raised dot in a given position in an isolated cell of Braille. Nonetheless, when attempting to read Braille normally, she found that she could not extract enough information to determine what letters, and especially what words, were written. She likened her impairment to "having the fingers covered by thick gloves". Despite her profound inability to read, she was struck by the fact that she did not notice any similar impairment in touch discrimination when trying to identify the roughness of a surface or locate items on a board. She was able to identify her house-key by touch in order to ask a friend to check on her cat at home, and she was similarly able to identify different coins tactually without any difficulty.

A subsequent neurological examination revealed no signs of peripheral sensory impairment in the hands and sensory and motor conduction velocities in the upper extremities were normal. Sensory evoked potentials from the right and left median nerve were unremarkable. An MRI of her brain (see Fig. 1), revealed extensive bilateral occipital lesions of increased T2 signal, most suggestive of bilateral occipital ischemic strokes.

Twelve months after her stroke she continued to be unable to read Braille and had resorted to the use of a computer with voice recognition software, otherwise she remained active and continued to work at the radio station.

## DISCUSSION

This patient became alexic for Braille following bilateral occipital strokes, probably secondary to an embolic stroke



**Fig. 1.** T2-weighted MRI of a patient who was an early blind formerly proficient Braille reader. Following her cerebrovascular accident, the patient developed Braille alexia.

at the tip of the basilar artery. Her MRI failed to document any lesions of the sensory motor cortices or to their projections. Similarly, imaging of her brain did not reveal damage to language centers, the angular gyrus, or white matter pathways leading to it. Peripheral motor and sensory nerve functions, as well as the sensory afferent pathways, appeared to be normal as documented by the neurological examination and neurophysiological studies. There was no discernible difficulty in performing non-spatial tactile perception tasks. The patient's impairment of tactile information processing was apparently limited to her Braille reading skill. Unfortunately, more detailed testing was not possible because of the patient's unwillingness to undergo further examination. Nonetheless, the findings strongly support the notion that in this early blind patient, the occipital cortex was indeed responsible for the decoding of spatial and tactile information required for Braille reading skill. This lends support to the experimental results obtained with rTMS of the occipital cortex in early blind subjects [2].

The network involving the striate cortex for haptic processing in blind Braille readers is thought to involve one of two circuits. Thalamic somatosensory nuclei may send outputs via projections to both the somatosensory and occipital cortices from multimodal cells located in the geniculate nuclei. Murata *et al.* [4] described the existence of these weak non-visual input cells to the striate in sighted, newborn cats and Berman [5] demonstrated an increase in thalamo-cortical connections to the primary visual cortex in cats following early removal of retinal input. Further evidence indicates, however, that cortico-cortico connections probably mediate the tactile perception of Braille in blind readers [6]. In monkeys, area 7 (posterior parietal cortex) has reciprocal connections to area 19 (visual association cortex) and visual deprivation in monkeys makes neurons in both areas responsive to somatic exploration [7]. This suggests that cortico-cortico networks

typically reserved for visual processing of object and pattern discrimination may eventuate in haptic discriminations in blind Braille readers.

Not all evidence gathered from early blind subjects implicates use of the striate cortex by blind Braille readers. The recent H<sub>2</sub><sup>15</sup>O-PET findings of Büchel *et al.* [3] differ substantially from the previous findings of Sadato *et al.* [1]. The Büchel *et al.* study reported that congenitally blind subjects (early blind subjects) show task-specific activation of extrastriate visual areas and parietal association areas during Braille reading, compared with auditory word processing, without activation of the primary visual cortex. In contrast, blind subjects who lost their sight after puberty (late blind subjects) did show additional activation in the primary visual cortex. Büchel *et al.* suggest that differential activation in late and congenitally blind subjects may be due to reciprocal activation by visual imagery in subjects with early visual experiences. Cohen *et al.*, on the other hand, have recently confirmed and elaborated upon the lack of involvement of striate and peristriate cortex in tactile Braille reading in late blind subjects. They suggest the possibility of a critical period of susceptibility for the recruitment of the occipital cortex for haptic information processing [8]. In any case, the findings in the present patient document the critical role played by the occipital cortex tactile Braille reading, at least in some subjects. It is plausible that the current patient's inability to read Braille stems solely from involvement of prestriate regions of the occipital cortex (e.g. Brodmann area 19). The location and extent of her occipital lesions are consistent, however, with bilateral involvement of the striate cortex (Brodmann area 17) in accord with the findings of Sadato *et al.* [1].

Alexia in Braille readers has only been reported in a few cases, and documentation of the underlying brain lesions is, in general, incomplete. Gloning *et al.* [9] studied a patient with a right parietal tumor who developed Braille alexia. The patient, however, died shortly thereafter and multifocal ischemic brain damage was found at neuropathological study. More recently, Birchmeier [10] studied a congenitally blind subject who, as the result of an ischemic attack, acquired an aphasic dyslexia for Braille, along with transient right hemiparesis. This patient had been an expert Braille reader prior to his illness. Unfortunately, any efforts that may have been made to localize the region of ischemic injury leading to the subject's specific impairments were not documented. Perrier *et al.* [11] reported the case of a patient who developed a severe Braille reading alexia without loss of verbal language skills following right hemispheric damage. The area of lesion, while not very well described, was extensive in both the parietal and occipital lobes, and Perrier and colleagues suggest that the patient's inability to read Braille might be related to a right hemispheric specialization for tactile information processing.

Finally, Signoret *et al.* [12] described a 77-year-old early blind individual who presented with Wernicke's aphasia, anomia, difficulty with word repetition and comprehension, and alexia with agraphia in Braille following an infarction of the left middle cerebral artery involving the temporal and inferior parietal lobes. A CT of the patient's brain revealed a lesion affecting Wernicke's area, the supramarginal gyrus, and the anterior portion of the

angular gyrus. The presence of alexia and agraphia for Braille in such a patient is not unusual in light of the location and extent of the lesion. In sighted persons, alexia with agraphia is typically associated with lesions of the angular or supramarginal gyrus of the parietal-temporal-occipital association cortex, since patients with these lesions are unable to connect visual symbols (letters) with the sounds they represent [13,14]. They are also unable to recognize embossed letters by touch, presumably because the angular and supramarginal gyri mediate the transfer of cutaneous sensory information into language areas [15]. Such a lesion interrupting the transfer of information between tactile sensory areas and language areas would readily explain alexia and agraphia for Braille in a blind individual.

By contrast, a lesion to the left visual cortex combined with damage to the splenium of the corpus callosum in sighted patients is associated with alexia without agraphia, since visual information obtained in the undamaged right occipital lobe cannot be conveyed to the angular gyrus and language areas of the left hemisphere [16]. Although no prior cases of 'pure' Braille alexia have been reported, analogous lesions in a blind Braille reader might presumably result in a pure alexia in a person who recruits his or her occipital lobes for the task of Braille reading. Although we do not know whether the present patient's Braille alexia was accompanied by Braille agraphia, we do know that her lesion is isolated to the occipital lobes, without the damage to the angular gyrus that one would expect in alexia with agraphia or the damage to the splenium that one would anticipate in a pure alexia.

## CONCLUSION

Because we could not perform more extensive testing on the patient, it is not possible to know for certain whether the patient's predominant symptom was an inability to make sense of the Braille symbols she could feel or whether she had simply lost the tactile acuity required to detect Braille stimuli. However, because she could detect other tactile stimuli, we believe that she suffered from a true alexia and that her perception of the stimuli was altered. Furthermore, because her language abilities other than Braille reading were intact, it appears that her language deficit was limited to perception of haptic symbols which mirrors previously reported cases of alexia in blind Braille readers. Overall, we interpret the findings in this patient as further evidence that Braille reading in the early blind is an example of cross-modal plasticity by which the occipital cortex is recruited for demanding tactile tasks, thus enhancing the sensory discrimination of blind subjects and making Braille reading possible.

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