Visual and Linguistic Impairment in Alexia Without Agraphia

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The classic syndrome of alexia without agraphia, first described by Dejerine as "pure word blindness" in 1892, has been repeatedly confirmed since then (Geschwind, 1962). Although diverse signs accompany the major symptoms, the constant features are inability to read printed material with retained ability to write both to dictation and spontaneously. Quite striking is the fact that the patient cannot read what he has written. Associated deficits which may appear in conjunction with alexia in the absence of agraphia include right homonymous hemianopsia (Benson and Geschwind, 1969), color agnosia (Geschwind and Fusillo, 1964), loss of ability to read musical notation (Geschwind, 1967), ability to read Arabic but not Roman numerals (Symonds, 1953), and anomia (Mohr, Leicester, Stoddard and Sidman, 1971).

Etiology of most reported cases has been vascular, specifically thrombosis involving the left posterior cerebral artery (Benson, Brown and Tomlinson, 1971). Few well-documented cases of this type of alexia resulting from trauma have been recorded, probably because the exact combination of lesions results from destruction of limited cerebral areas. Onset is typically sudden and often appears without paralysis, sensory loss or significant aphasia. Destruction of commissural pathways from the right visual cortex to the dominant language center is the suggested anatomical basis. A lesion in the splenium of the corpus callosum prevents visual language information from the right hemisphere from reaching the intact left supramarginal gyrus (Geschwind, 1965). As a result, the intact right visual cortex correctly perceives visual stimuli as words but the callosal lesion prevents these images from "re-auditorization" into graphic symbols.

Although a great number of cases have been reported in the literature, it is uncertain to what extent the alexic impairment is of a visual or a linguistic nature. Various researchers have distinguished between literal and verbal alexia (Luria, 1970; Benson, Brown and Tomlinson, 1971), Stage I (sensory) and Stage II (perceptual) disorders (Brown, 1974), or surface and deep alexia (Marshall and Newcombe, 1971; Gardner, 1974). To clarify terms used in this paper, visual impairment in alexia is seen in the inability to clearly perceive letters as graphic symbols or confusions of graphemes with similar visual configurations. On the other hand, difficulty decoding linguistic information (words or sentences) is apparent when errors bear a semantic or grammatical relationship to the stimulus item and are usually designated as paralexic. Since most "pure" alexics have difficulty with both processes, visual and linguistic alexia are merely points on the continuum. By carefully examining the patterns of alexic impairment in a single patient, it is hoped that more thorough evaluation and realistic rehabilitation of the patient who presents alexia without agraphia can be undertaken.

The objective of this study was to explore perceptual and linguistic variables that may affect performance when alexia is the primary feature of language impairment. In the study of this patient, we were influenced by our conviction that the structural types of errors performed provide one of the most important constraints upon theories of reading impairment. We also wanted to call into question the widespread assumption that reading is a single capacity which can break down totally.

CASE REPORT

Clinical History: The patient studied, an 81-year-old, college educated, white female, experienced loss of memory and confusion one evening 5 months prior to this investigation. On admission to the hospital, it was determined that the patient had suffered a cerebrovascular accident resulting in aphasia, right homonymous hemianopsia and mild right hemiparesis. Brain scan was normal. Electroencephalogram revealed left parietal lobe infarction. On discharge ten days later, the patient showed only alexia and partial right homonymous heminopsia. Her physical condition subsequently stabilized and no further signs of neurologic disease have appeared.

Procedures

Investigation of this patient consisted of conventional language testing and a number of special evaluation procedures. All were conducted in five sessions at her retirement home. No formal treatment was in progress during this investigation.

General Language Testing

The Minnesota Test for Differential Diagnosis of Aphasia (Schuell, 1965) was administered while the patient was hospitalized. Four months later, the Minnesota, supplemented by portions of the Boston Diagnostic Aphasia Examination (Goodglass and Kaplan, 1972), was administered by the present authors.

Results: Findings of both examinations were compatible with the original diagnosis of alexia with mild anomia. Auditory recognition and retention span were undisturbed; seven digits were repeated forward without difficulty. Although auditory comprehension for carrying out directions was unimpaired, some difficulty in remembering information read to the patient was noted. Mild anomia characterized verbal expression. The patient was able to name pictures of common objects and actions and to provide names of concrete objects in her environment. However, in conversational speech, she would occasionally search for words, particularly proper nouns. Reading of single words, especially those with similar visual configurations, e.g., house and horse, was difficult. The patient was unable to comprehend sentences or paragraphs when presented visually. She stated that although the words looked familiar, she was unable to glean the meaning. Greater difficulty in copying written language and geometric figures than in self-generated graphic responses was noted. Both oral and written spelling were intact. Arithmetic calculations were restricted to simple addition and subtraction.

Reading

To ascertain the nature of the reading impairment, a battery of tests was given. It consisted of two types of tests, each being prompted by the hypothesis proposed, i.e., alexia may be underlined by primarily visual problems (so called surface alexia) or it may be underlined by primarily linguistic problems affecting the structure of grammar and lexicon (so called deep alexia) or the combination of both types.

The following tests were given to explore surface alexia:

1. Reading of letters in various prints, Arabic and Roman numbers and conventional symbols, e.g., a stop sign or dollar sign.
2. Short as opposed to long and/or visually complex words, e.g., cross; fingernail; through.
3. High frequency as opposed to low frequency words, e.g., letter; prosecutor.
4. Pairs of visually similar words, e.g., bear; pear.
5. Misspelled words, e.g., appland for applaud.
6. Words familiar to the patient as opposed to unfamiliar, e.g., Texas; atomic.

The tests for exploring deep alexia involved four categories of words:

1. Words involving rules of grapheme-phoneme correspondence.
   a) final /gh/words, e.g., rough, although.
   b) silent grapheme words, e.g., glisten where the /t/ is silent.
   c) vowel name words for possible contamination of the reading process by lengthening the sound of the vowel, e.g., glad may be read as glade.
2. Words exhibiting various degrees of morphological complexity in terms of number and types of morphemes.
   a) -er words consisting of single morphemes, e.g., finger.
   b) -er words consisting of two morphemes, e.g., teacher.
   c) -er words exhibiting noun/verb ambiguity, e.g., master.
   d) words exhibiting derivational complexity, e.g., impossibility, aggressiveness.
3. Words belonging to various form classes.
   a) function words as opposed to content words, e.g., after, wall.
   b) nouns as opposed to verbs and adjectives, e.g., give, take, red.
4. Words exhibiting specific semantic relationships including:
   a) kinship terms, e.g., father, sister.
   b) Antonyms, e.g., wide - narrow; big - small.

In addition to the above word-reading tests, phrase and sentence reading tests were also administered. They consisted of:

1. Familiar phrases and associations, e.g., salt and pepper; up and down.
2. Low frequency phrases and sentences, e.g., The phantom soared across the foggy heath, taken from the Boston Exam (Goodglass and Kaplan, 1972).
3. Reading and matching sentences with appropriate pictures. For this task the Northwestern Syntax Screening Test (Lee, 1969), presented visually, was used.

Results: The patient exhibited problems associated with visual or surface alexia, as marked by the types of errors described below. Manifestations of this problem as described by the patient included words changing as she looked at them, looking funny, either too long or too short for the given words. Also the inability to differentiate certain graphemes and difficulty in seeing two identical graphemes in sequence, e.g., pretty, were observed. Difficulty in shifting from one line to another while reading was reported. Size of print affected the patient's performance, in that bigger print was difficult for her to read, according to her own report. (This finding is in direct conflict with that reported by Woods and Poppel (1974).)

In general, her reading performance can be characterized in the following points:

1. Reading impairment was manifested by failures to read, misreadings, delays and resorting to spelling the word out (either vocally or subvocally) when in difficulty.
2. Reading was performed by attending to individual graphemes sequentially and no ideographic reading was observed. Reading was characterized by incomplete visual analysis of the stimulus.
3. Misreadings and misspellings were consistent.
4. Very few neologisms were produced. Those produced resulted from grapheme confusions, e.g., through for thorough.
5. Misreadings corresponded to the length and syllabic structure of the target word.
6. Misreadings involved primarily endings of words.
7. In morphologically complex words, misreadings observed morphemic boundaries. Since the misread morpheme was usually the last one, there was a tendency to omit the morpheme, which resulted in morphological simplification, e.g., plural nouns became singular, past tense verbs became present.
8. The number of misreadings and latency were related to the length, visual complexity and the patient's familiarity with the word.
9. No disturbance of grapheme-phoneme correspondence rules was found.
10. No effect of grammatical or semantic complexity was observed in the errors performed in reading words.
11. Only in sentence reading, both semantic and grammatical substitutions occurred.
12. Paragraph reading was not possible for this patient.

The specific error types were associated with the visual recognition of the shape of the individual graphemes. They consisted of:

1. Overall gestalt confusions, e.g., /k/ for /r/, speak for spear; /g/ for /r/, message for massacre.
2. Reversal errors, e.g., /b/ for /d/, rob for rod.
3. Inversion errors, e.g., /b/ for /p/; /u/ for /n/; /d/ for /g/, applaud for appland.
4. Word-final omissions, e.g., glasses - glass; slippery - slipper.
5. Total inversion, e.g., comb for bone.

Number Reading: No differential impairment in reading of selected Arabic as opposed to Roman numerals was present. However, some delays and self-corrected responses were observed.

Writing

Letters, Arabic and Roman numerals, single words and sentences were dictated for the patient's response. A picture was used as stimulus for a descriptive paragraph. Single letters, short words and sentences written in block letters were presented for copying.

Results: Writing to dictation revealed only minor distortions of letters (Figure 1). Writing of a paragraph when self-generated was within normal limits (Figure 2). Copied writing was greatly altered. Mirror image reversals and axial rotations were observed in copying upper and lower case block letters (Figures 3, 4, 5). In addition, paragraphic responses, e.g., planter for planted, were observed in copying sentences (Figure 6). In both

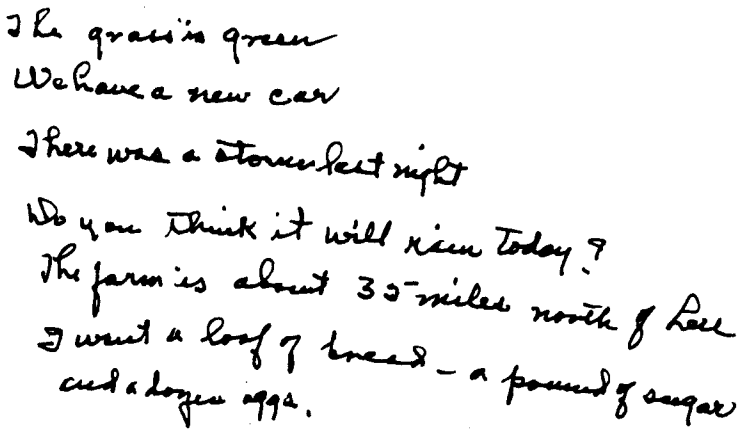

Figure 1. Writing to dictation.

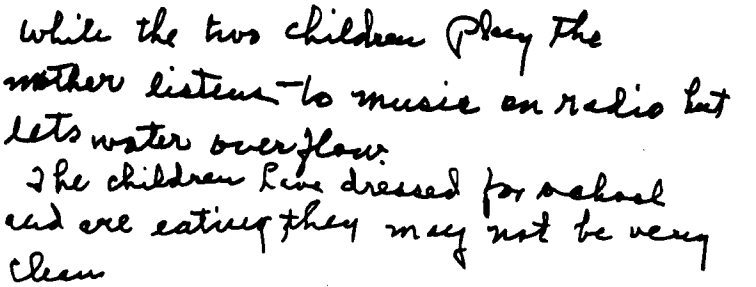

Figure 2. Writing a self-generated paragraph.

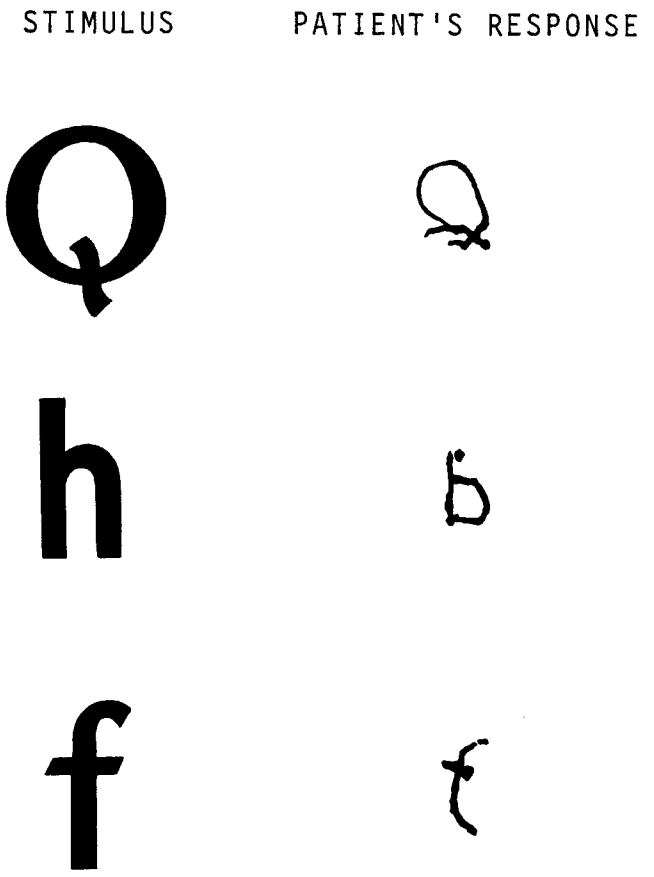

Figure 3. Copying letters.

STIMULUS PATIENT'S RESPONSE

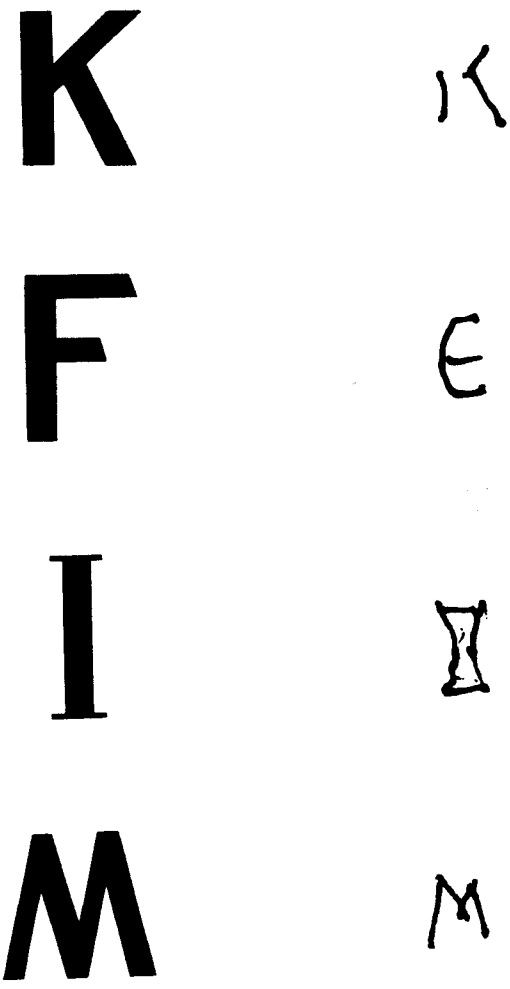

Figure 4. Copying letters.

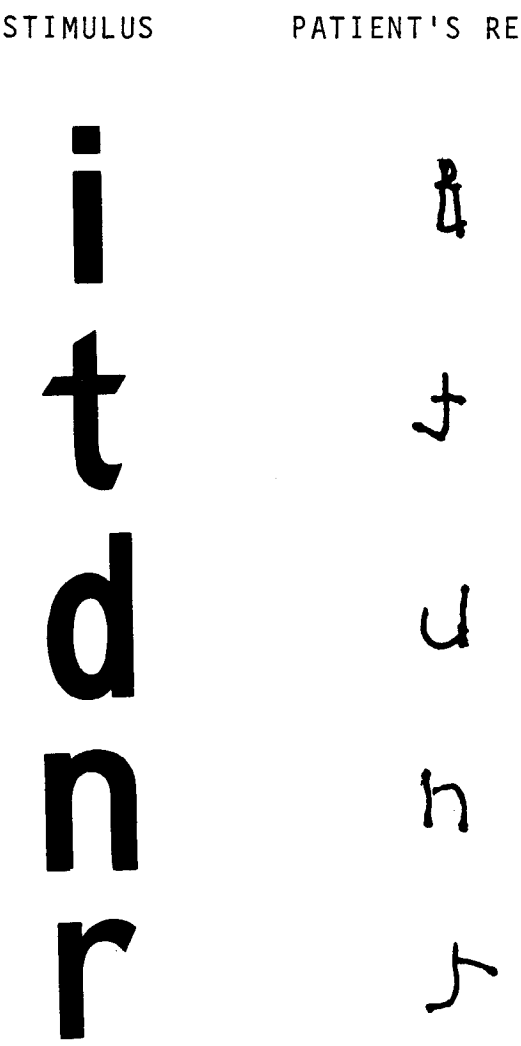

Figure 5. Copying letters.

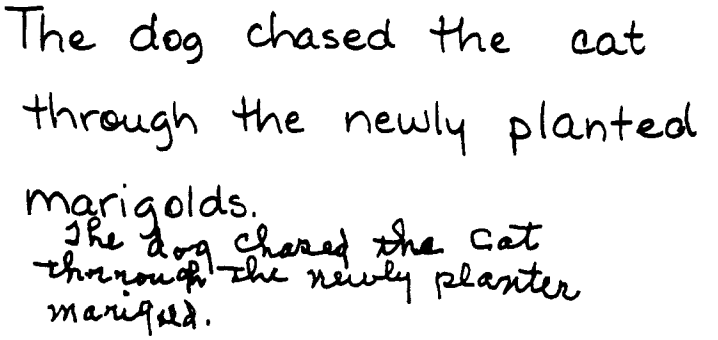

Figure 6. Copying sentences.

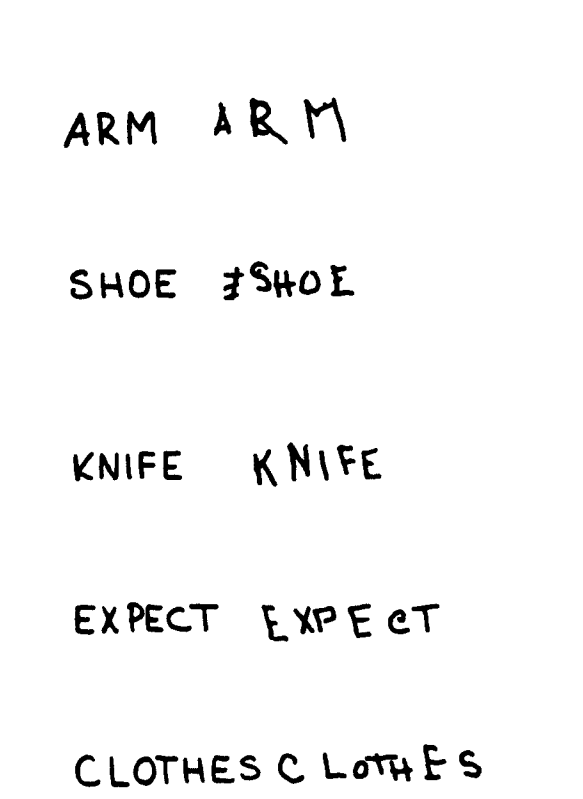

Figure 7. Copying words.

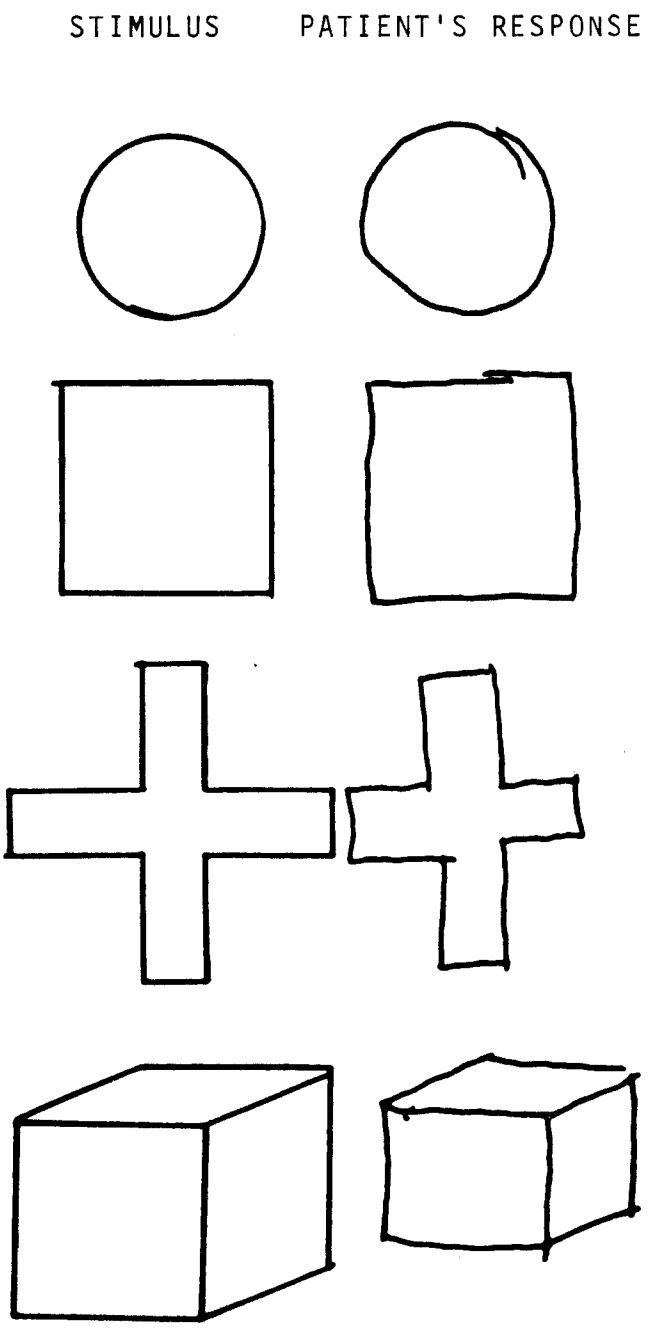

Figure 8. Copying geometric figures.

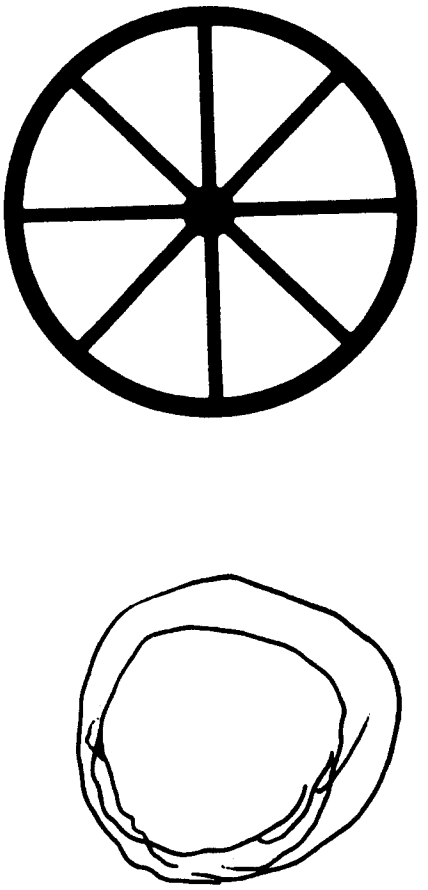

Figure 9. Copying a wheel.

Figure 10. Drawing a man.

reproducing printed upper and lower case letters from visual memory and writing them to dictation, distortions were present. More difficulty was encountered with block letters than cursive letters. Copying short words and sentences in block letters also revealed distortions (Figure 7). However, in writing words or sentences to dictation or composing a short paragraph, fewer of these distortions were present than in copying. Difficulty moving from one line to the next and a downward slant to the right characterized her writing.

Drawing

Copying of two- and three-dimensional figures was within normal limits (Figure 8). In drawing a picture of the wheel from a model, the production was impaired in that no spokes were included in the patient's drawing although she recognized the missing components (Figure 9). In drawing a man from memory, an impressionistic sketch was produced. No facial features were present. Closer examination revealed short, unconnected lines suggesting only major parts of the body (Figure 10).

Color Identification

A number of tasks were presented to the patient to evaluate her color perception ability. For color naming, crayons and colored pictures were used. Both pictures of objects with strong color associations, e.g., a lemon, and those of arbitrary colors, e.g., a dress, were presented for matching to a color chip. Three series of varying shades within a hue were given for arrangement from the lightest to darkest shade. Writing the color name of an object commonly associated with the color, e.g., a bride's dress being white, completed tasks of color identification.

Results: Results of testing color perception in this patient revealed that she had difficulty in the categorical perception of colors. The patient described colors in terms of hue differentiation using the "-ish" form of the adjective, e.g., "pinkish, but I don't know what name you'd call it." Difficulty distinguishing black, brown and gray was observed. In matching a color to an artibrarily colored picture, the patient accepted only colors identical to the hue of those pictured and not representative of the category. However, perception of shades within a hue was intact as shown by her ability to arrange chips in terms of intensity.

Color naming performance also reflected impairment in categorical perception of color. Of the ten crayons presented, only two, green and purple, were correctly identified. When asked to write the color of an object commonly associated with that color, the patient frequently wrote the entire association, e.g., "green grass" or "egg yolk--pale yellow." Otherwise graphic naming of colors was without error.

DISCUSSION

Comparing these clinical findings to those described in the literature, it may be speculated that the patient studied exhibits a callosal lesion which disconnects the visual cortex of the right hemisphere from the language areas of the left hemisphere. The separation of these areas not only produces alexia but deficits in color identification. Both processes involve visual-auditory associations which are essentially void of somoesthetic associations (Geschwind and Fusillo, 1964). Reading and color-naming are learned primarily through visual-auditory associations while naming of objects and numbers is learned with considerable somoesthetic reinforcement. Objects may be handled and numbers counted on the fingers but words and colors provide no tactile or proprioceptive stimuli. Infarction in the territory of the dominant

posterior cerebral artery has also been associated with the particular type of anomia described (Mohr, Leicester, Stoddard and Sidman, 1971). It may be that this patient's anomia which is limited to names of people and places is an extension of the visual deficit. These items are as intangible as colors and graphemes.

Further evidence of visual disturbance was noted. Both in reading and writing, the patient showed difficulty in moving from one line to the following. She described the lines and words as "jumbled" or moving while she looked at them. Copying was disturbed, with self-generated written responses being superior.

Cursory examination of reading impairment generally performed as part of an aphasia test battery does not allow for in-depth evaluation of alexia. As our investigation provides a closer examination of the nature of the alexic deficits, we would like to postulate that visual or surface alexia may occur in isolation and leave the grammatical and semantic features of words relatively intact. However, even in cases of primarily visual deficit, the data must be integrated with the studies of the relative power of different word-fragments (initial, medial, and final sequences) and contrasts due to morphological structure (inflectional and derivational affixes) which influence the "chunking" of visually presented words.

Clearly recognition of the type of impairment, whether visual or linguistic, has much to do with realistic remediation of the problem. If the problem concerns deeper structures of language, not only alexia involving the syntacticosemantic system, but other aphasic features are present. Reading becomes secondary to the more widespread language problem. When impairment is primarily visual, only surface deficits are seen and the phenomenon may be strictly isolated from other language problems. As clinicians, we recognize that language disturbance is rarely an all or none process; error patterns observed can be related to the hierarchy proposed.

Implications for treatment: While it is not possible to conduct formal treatment with this patient, examination of the literature regarding prognosis of alexia without agraphia and of the changes in the patient's behavior during the five-week period of testing, provide implications for treatment. Prognosis in alexia is an unsettled question, and limited reports of rehabilitative measures used with this particular syndrome have been published. Ajax (1967) in an extensive report of retraining techniques used with two patients, concluded that the prognosis seems no better than in more extensive disorders of language. Other investigators (Newcombe, Marshall, Carrivick and Hiorns, 1974; Thomsen and Harmsen, 1968; Ulatowska and Richardson, 1974) speak of recovery in acquired alexia.

Although the patient reported in this case study did not receive any formal treatment, her reading did improve. She exhibited the desire to read and attempted to read magazines daily. It is suggested that the extensive testing of her reading as described previously formed the basis for improvement. Since her linguistic competence was well preserved, techniques of direct confrontation of errors were possible. Mistakes in visual decoding were pointed out and the patient was encouraged to correct her responses. As testing progressed, it was possible to demonstrate rules of grapheme/phoneme correspondence. Because sight word methods which sometimes failed appeared to be the basis for her analysis of words, morphological structure of words was explained. This method helped her read words of greater length and morphological complexity.

Acknowledgement

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DISCUSSION

Did you test your patient's ability to use tactile and proprioceptive input, i. e., tracing? We have found with two patients having this syndrome (in Memphis) that if you give them short words in very large print; they do very well, and that they can identify letters very accurately. We assume that they achieve it through eye muscle tracing of some sort. They can also do it by tracing in their hands. I wondered if you have tested that?

We have not done tracing testing. It is unfortunate that we do not have the data to compare with yours. We have found in relation to the size of print was very striking and in direct conflict to what has been reported in the literature, i.e., the bigger the print, the better the patient's reading abilities. That did not happen with our patient; she complained that she could not read large print, and we were forced to go to smaller size print for her to read.

Further, what was so surprising to me in this patient is that as we tested, she improved dramatically. She had been this way for five months with little improvement. She worked very hard every day, reading magazines on her own. She was highly motivated but there was no therapy available to her. What we were doing was "diagnostic therapy."

Since the patient reported that bigger print was more difficult did you test her reading skills on that print before moving to smaller print?

Yes, we tested her on both and observed delays in performance on the former.

I wondered if she looked at this as too immature, too primary in level and was therefore objectionable to her.

We do not feel that this was the case, since her cooperativeness was high.

We have found that there are two different groups of patients. If the lesion is primarily between the visual systems and posterior intrinsic system, tactile cues and eye tracing help. But if the lesion is deeper in the parietal area, that does not help at all because the patient is getting the information all right, but it is a decoding problem. You are talking about two different groups of patients.

We found a characteristic test profile showing depression of auditory component and reading skills with relative preservation of graphic skills and anomia.

PICA testing was not possible. Frequent interruptions and lack of controlled atmosphere in this patient's retirement home prevented PICA testing. As an extension of the PICA profile just described, I would like to examine the slides of the patient's graphic responses (See examples and discussion in the body of the text.) Note the discrepancy of skills between self-generated writing and copying. We feel that distortions in copying are an extension of the reading disturbance.

Have you done any testing with visual imagery? We also have a patient with this rare syndrome. He complains of a deficit in visual imagery. He says that as long as he can image it, he can say it. Does your patient show

strong adaptations to her heminopsia? Do you see it in her walking? Our guy hugs the wall.

She recognizes the heminopsia and scans when reading. Otherwise, no unusual compensatory behavior was noted. We have not tested visual imagery.