Visual disorders of higher cortical function

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ABSTRACT: Following stroke or other causes of brain damage, patients may demonstrate visual disorders of higher cortical function. These defects involve visual attention, oculomotor skills, visuospatial orientation and object recognition. It is important for optometrists to understand these conditions and consider their presence in individuals with persistent visual complaints despite a normal exam, or in patients who fail to respond to magnification in low vision rehabilitation. The diagnosis and management of these higher order visual disturbances are discussed.

KEY WORDS: hemi-inattention, visual agnosia, alexia without agraphia, Balint's syndrome, higher order visual disturbances, rehabilitation, stroke, brain damage

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*1*Most optometrists are familiar with the myriad of visual perception disorders occurring in infants and children. They are trained to administer and interpret tests that detect these congenital or developmental disorders occurring most frequently in children with learning disability, attention deficit disorder, mental retardation. and cerebral palsy. 1 Within the optometric literature, little attention has been given to acquired visual disorders of higher cortical function that occur in the adult or elderly population as a result of brain damage from infarction, hemorrhage, trauma, tumor or senile dementia. It is the purpose of this paper to review these neurological syndromes of disturbed visual perception and to discuss the importance of their recognition.

Unilateral spatial inattention

Unilateral spatial inattention or neglect is the involuntary failure to attend to or respond to meaningful sensory stimuli presented in the affected hemispace.' This phenomenon may occur with any combination of visual, auditory and tactile stimuli. Although this is not caused by motor or sensory defects, it is frequently accompanied by hemianesthesias, hemiplegias and homonymous hemianopsias. The effect of hemispatial neglect can be devastating. In fact, some researchers cite unilateral neglect as one of the main factors interfering with recovery and successful rehabilitation in stroke patients' In severe forms, the patient demonstrates complete hemispatial neglect to the extent that he may not dress or shave the affected half of the body/ He may not recognize his arm and leg as his own and object to their presence in his bed." Questions addressed to the patient from the affected side may go unanswered and navigation may be difficult if the patient collides with objects and is unable to orient himself to new surroundings.* In its most subtle forms, the patient demonstrates inattention only when faced with a complex task. In these cases, the hemispatial inattention may be detected by presenting the patient with symmetrical simultaneous stimuli (i.e., fingers presented to each side of the mid-line in the superior visual field). Failure to detect both stimuli constitutes an abnormal response known as the extinction phenomenon.5.6

Hemispatial inattention is usually the product of a right (non-dominant) parietal lobe lesion. Left sided parietal lesions are less commonly reported and are associated with less persistant and milder forms of inattention."7 Rarely there have been other reported cases of lesions occurring elsewhere in the brain including the frontal lobes, occipitoparietal lobes, cingulate gyrus, thalamus, putamen, basal ganglia and mesencephalon.'J Infarction (stroke) is most commonly implicated, although the phenomenon has been reported in association with hemorrhage, tumor, and traumatic contusion.2 The exact mechanism is unclear.

The examiner's assessment should begin with simple observation of the patient. As mentioned earlier, patients with moderate to severe hemispatial neglect may demonstrate peculiar behavioral

patterns as they orient their body and activities toward the unaffected hemispace (ipailateral to the side of the lesion). For example, they may comb only half of their hair, fill out only half of the insurance form or read only half of the letters on each line of the acuity chart. The patient's family members or case workers may also be able to provide a history of this type of behavior.

A commonly used screening procedure for hemispatial visual inattention is the double simultaneous stimulation test, which was first described by Oppenheim using tactile stimuli.2 A standard confrontation visual field is first conducted using single stimuli presented in each quadrant of the visual field, one at a time. If the responses are abnormal (i.e., failure to identify the stimulus), one cannot differentiate between a hemianopic visual field defect and a severe attentional disorder without further testing.* In this case, the double stimulation test cannot be performed. If the responses are normal, however, the examiner then presents double simultaneous stimuli, placed on each side of the vertical midline. Extinction occurs when the patient fails to perceive the stimulus in the field contralateral to the lesion, while correctly perceiving the stimulus in the ipsilateral field. Although sensory extinction, like hemi-inattention, is typically a product of right parietal lobe lesions, it is not pathognomonic for hemi-inattention disorders." In addition, the test sensitivity may be as low as 50 percent 9

Pursuit and saccade testing are also easy screening procedures that may alert one to the presence of lesions that can produce hemispatial inattention. Affected patients may demonstrate difficulty when attempting to track an object into the affected hemispace and maintain stable fixation. When asked to alternate fixation between two targets on either side of the midline, they may fail to search or demonstrate very slow searching in the hemispace contralateral to the lesion. Optokinetic nystagmus (OKN) reflexes also may be of some benefit as they are typically decreased or absent when the drum is rotated towards the side of a parietal lobe lesion."

For patients with concurrent hemianopic or quadrantanopic visual field defects, the double simultaneous stimulation test cannot be performed, and pursuit/saccade testing may not produce reliable results since abnormal responses may occur as a result of the visual field defect alone. For these patients, the clock completion test and the line bisection tests are particularly useful.

In the clock completion test, the examiner draws a circle (clock face), and then asks the patient to complete the clock by drawing in the numbers and hands. The test material should be placed in the attended space, opposite the side of visual field loss. Patients with hemispatial visual inattention will either place all elements of the clock on the side ipsilateral to the lesion or they will complete only this half of the clock (Fig. 1). This assymetry in performance allows the examiner to differentiate this defect from other visuoperceptual or constructional difficulties, and placement of the test in the hemifield free of visual field defect reduces field loss as a cause of poor performance. Similarly, the examiner may draw a series of horizontal lines and ask the patient to draw an "X" at the center of each line." Patients with hemispatial visual neglect may fail to cross all the lines on the page, and of those lines which are crossed, the "bisecting line" is skewed toward the side of attended space. A visual marker (such as a letter) may be placed at the beginning and end of each line and the patient is then instructed to bisect the line only after naming the letters associated with each line. This ensures that the entire line is seen, thereby eliminating any effect from visual field deficits.

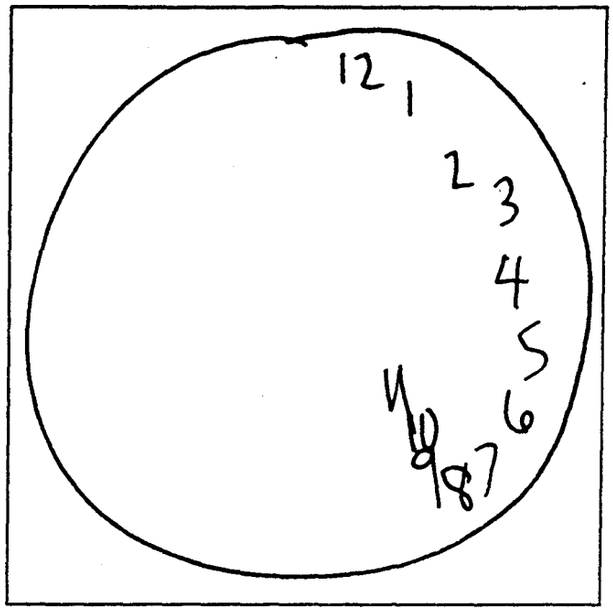

Figure 1: The results of the clock completion test, as performed by a person with hemispatial visual inattention.

The Rivermead Behavioral Inattention Test (RBIT) has been developed as a useful method for detecting visual neglect, as well as for estimating its functional impact.'3 The RBIT differs from other tests of visual inattention because it uses behavioral measures such as eating a meal, reading a menu, etc. ~

Rehabilitation of patients with bemispatial visual inattention is difficult. A common sense approach to training, aa outlined by Bouska, et al., is found in Table 1.' One should keep in mind that although patients may be trained for specific tasks, they may find it hard to generalize and apply these techniques to untrained tasks' In addition, some patients manifest other cog-

Table I: Training guidelines for hemispatia visual inattention

1. Attempt to improve patient's awareness and understanding of inattention and/or visual field loss using examples in the environment.
2. Practice visual scanning skills, including ocular pursuit and saccades, into the unattended space.
3. Provide the patient with increasingly complex visual-perceptual and visual-motor tasks which require the integration of the above two points.
4. Stress the importance of scanning during activities of daily living-, reminding the patient, "when something doesn't make sense, look into the unattended space and it usually will".
5. Use visual markers at the end/beginning of the page to decrease inattention while reading.

nitive, motor or sensory deficits that may complicate or prohibit attempts at rehabilitation.

Visual agnosia

This term is used to describe a number of disorders characterized by the inability to recognize an object by sight, despite adequate cognition, language skills and visual acuity/field." In its pure form, object recognition is achieved only by auditory, tactile or olfactory stimuli. As an example, a patient with visual object agnosia is shown a set of keys on a ring. Although his vision is good enough for him to accurately sketch the keys, he is not able to verbally identify them, describe their use or signal his recognition by demonstrating their use. However, by holding the keys or hearing them jingle, the patient is able to correctly identify them as a set of keys.

Since the first description by Lissauer in 1889, the visual agnosias have been classified into two groups; the associative form and the apperceptive form." In order to best understand these terms, recall that the eye, the optic nerve, optic tracts and the occipital cortex are responsible for generating the primary visual input. These signals must undergo some form of intermediate processing prior to the mind's recognition of the object. If the pathway is damaged immediately after the most elementary sensory level, then a normal percept is not formed, resulting in an apperceptive agnosia. Damage occurring after the intermediate processing results in the inability to integrate the perceptual information with the stored knowledge of the object and its functions. This classification scheme is, at best, incomplete. Numerous case reports do not fit well into either category, suggesting the need for an expanded system."" Some authors argue the existence of this dichotomy

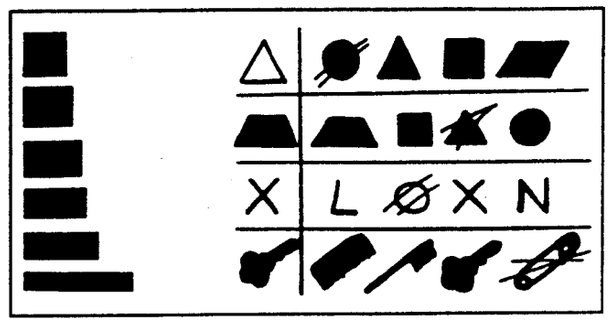

Figure 2: Patient with apperceptive agnosia demostrating inability to correctly match object shapes. (Drawing reproduced with permission from Farah MJ, Visual Agnosia. Cambridge, Massachusetts: The MIT press, 1990.)

altogether, claiming that all patients with visual agnosias would show significant visual pereeptual difficulties if subjected to adequate study."" For the sake of this discussion, however, we will continue to follow this classification scheme.

Associative agnosia

As mentioned previously, visual perception is adequate, however, the percept is "stripped of its meaning" because it is separated (disconnected) from the patient's memory and associated knowledge of the object's functions.'g The most common types of associative agnosia are object agnosia and prosopagnosia. Less common forms include color agnosia and topographic disorientation.

Associative visual agnosias: object agnosia

Patients with the associative form of object agnosia are unable to recognize objects by visual inspection alone. They are not able to name the object, describe its use or demonstrate its use. They are also not able to sort objects into different groups based on similar functions (i.e., kitchen utensils and garden tools). If the examiner names one object in a group of objects, the patient is not able to identify the correct object, however, if the patient is shown the same object, he is able to match it to the identical object in the sample group. The patient may be able to sort unrecognized objects into visually similar groups (i.e., sphere shaped apples and long skinny pencils). He is also able to copy or draw the object in such a way that it is recognizable to others (Fig. 2). Object recognition may be achieved only by touching, smelling or hearing the object.

There are varying degrees of difficulty in visual recognition, thus many patients do not exhibit complete object agnosia. Starting with the easiest, progressing to the most difficult, classes of visual recognition tasks include: real items in use, real items not in use,, photo-

graphs, line drawings, and complex pictures.2011 In addition, we know that recognition performance is decreased if part of the object is obscured or if the object is presented tachistoscopically."-"

It is important to distinguish object agnosia from optic aphasia. The latter is a language disorder in which the affected patient cannot name visually presented objects, but he can signal his recognition by pantomiming their use, or by pointing to objects named by the examiner.

Although the exact mechanism is not known, it is clear that most cases of associative object agnosia result from bilateral lesions in the occipitotemporal lobes with damage to the cortex and subjacent white matter.~M Infarct is most commonly implicated as this region is supplied by basilar artery circulation." Other causes may include tumors, carbon monoxide poisoning and Aizheimer's disease."'~ There have been a few reported cases of associative object agnosia caused by unilateral damage to the occipital lobes at the junction of the posterior temporal or parietal lobes, either on the right or left sides.'*

Other clinical findings commonly associated with object agnosia include visual field defects (unilateral or bilateral homonymous hemianopsias or quadrantanopsias), prosopagnosia, alexia, topographic disorientation and achromatopsia.20·25

Associative visual agnosias: prosopagnosia

Prosopagnosia is the inability to recognize faces despite unimpaired visual recognition of most other stimuli. These patients have the most difficulty with similar appearing faces such as family members, and they may not even recognize their own face in the mirror. Recognition is achieved when the person speaks, or the patient may learn to "key" their recognition of a person by the shape of the hair line, a mustache, or side***b***U*m*s.20.26 This defect of recognition is not confined to human faces, rather it often extends to involve similar objects within the same class.~" Affected patients may not be able to distinguish between types of dogs or types of birds. Similarly, they may not be able to tell the difference between types of plants, makes of automobiles, styles of chairs or even types of clothing."" As an example, a patient with prosopagnosia may be able to generically identify an object as a car, but he may fail to identify his own car in a parking lot full of others. Although prosopagnosia is often classified as a type of associative agnosia, controversy exists regarding the integrity of the percept formed. There is some evidence that at least some patients do not perceive the faces normally."

Screening for the presence of prosopagnosia may be informally conducted by asking the patient to identify photographs of famous people and/or family members.

As with object agnosia, the exact mechanism is uncertain, and the lesions are usually bilateral in the inferior occipitotemporal *regions.'"]0* It is felt by some that involvement of the inferior longitudinal fasiculus is a prerequisite for the occurrence ofprosopagnosia.30·3"

Associative visual agnosias: achromatopsia and color agnosia

Patients with cerebral achromatopsia demonstrate preserved form vision, but because of a loss of color vision, are unable to discriminate between different colors or hues.20 Total achromatopsia is clearly evident to the patient as it causes the entire world to appear in shades of grey. They are not able to name the colors of cards fully saturated with color, they are not able to point to the color named by the examiner and they are not able to sort colors into groups of basic colors. They are unable to correctly identify any plates on the pseudoisochromatic tests of color vision. Partial achromatopsia may occur, affecting only a hemifield or a portion of the hemifield in which case the patient may not even be aware of the deficit. Superior visual field defects on the affected hemifield, as well as prosopagnosia and object agnosia may be associated findings.* Lesions in the inferior occipital lobe or occipitotemporal region are usually responsible for producing central achromatopsia.

Color agnosia is evident when the patient is unable to name the color or point to the correct color when given a color name. They are, however, able to sort and match colors, and they score normally on the pseudoisochromatic tests of color vi***s***ion.33 Color agnosia is commonly associated with alexia, and is usually produced by lesions occurring in the left occipital lobe and the splenium of the corpus collosum.32

Associative visual agnosias: environmental agnosia

Topographic disorientation or environmental agnosia is the inability to recognize the features or landmarks of a familiar place or a region. This disorientation precludes successful navigation around town, in the hospital, and even in the patient's own home in some cases. The patient may be able to compensate to some extent by using verbal directions such as "the kitchen is two doors past the bathroom." Formal testing for topographic disorientation is rarely needed as this disorder is quite apparent as the patient attempts to carry out his activities of daily living.

Environmental agnosia is usually produced by lesions in the right medial occipitotemporal region." It may occur in association with left hemianopic visual

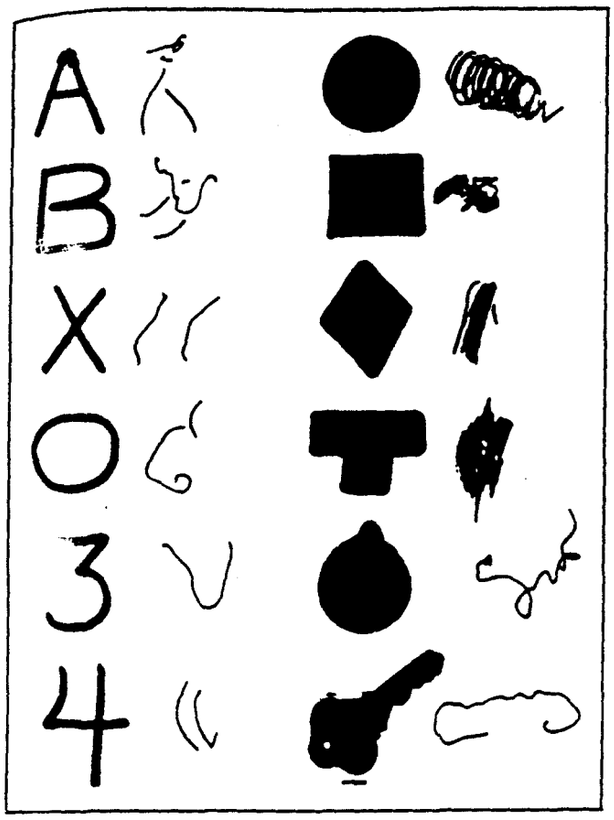

Figure 3: Same patient as in Picture 2, demonstrating inability to copy simple figures as a result of apperceptive agaosia. (Drawing reproduced with permission from Farah MJ, Visual Agnosia. Cambridge, Massachusetts: The MIT press. 1990.)

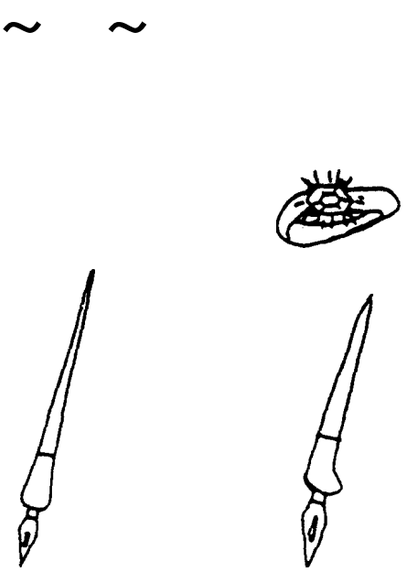

Figure 4: Patient with associative agnosia who could accurately reproduce line drawings, but was unable to recognize the objects. (Drawing reproduced with permission from Farah MJ, Visual Agnosia. Cambridge, Massachusetts: The MIT press, 1990.)

Table 2: Diagnostic features in the classification of associative and apperceptive agnosias

Functions to be tested:	Associative	Apperceptive
1. name object	No	No
2. match identical objects	Yes	No
3. describe object shape	Yes	No
4. sort by similar shapes	Yes	No
5. sort by similar functions	No	No
6. draw/copy object	Yes	No
7. pantomime use of object	No	No
8. identify object via touch, sound or feel	Yes	Yes

field defects as well as visual hemineglect. Although the latter disorders may contribute co failure at some travel-related tasks, they do not produce the significant degree of disorientation observed in environmental agnosia.

perceptive agnosia

In contrast with associative visual object agnosia, the patient with apperceptive agnosia is unable to match similar shapes or accurately draw/copy objects due to inadequate visual perception (Figs. 3 and 4). As with associative agnosia, recognition is achieved only by auditory, tactile or olfactory stimuli (Table 2). Documented cases of apperceptive agnosia are rarely re-p'-ted and are most often attributed to more diffuse cerebral damage such as that from anoxia or carbon monoxide poisoning. The brain damage usually affects the occipital lobes and surrounding areas'

Rehabilitation techniques

Little information is available regarding the rehabilitation of patients with visual agnosias. If one assumes that patients can re-learn to recognize objects and people, a treatment plan can be developed that involves repetitive discrimination drills using objects, color cards and photographs. Likewise for topographical disorientation, the patient may be instructed to repetitively practice simple travel routes that become progressively more complicated." As with blind rehabilitation, the importance of teaching compensatory techniques should not be underestimated3'38 Family members should also be educated as to the functional implications of the disorder and should be made aware of compensatory techniques as well.

Alexia without agraphia

Alexia is characterized by the inability to comprehend written or printed material. It should be noted that this is an acquired disturbance of reading and should be distinguished from dyslexia, which is generally regarded as a developmental condition. Since Dejerine's landmark publications in 1891 and 1892, it has generally been accepted that two main types of alexia exist, depending on the presence or absence of a concurrent writing disturbance (agraphia). More recently, a third type of alexia has been described among patients exhibiting both reading disturbances (alexia) as well as speech/language disturbances (aphasia)."

In its purest form (alexia without agraphia), reading comprehension is severely disturbed despite intact spontaneous writing or writing to dictation. In addition, the patient demonstrates normal spelling and does not suffer from aphasia or dementia. Because the symptoms are dramatic and in the visual realm, these patients often present initially to their eye care practitioner.

The extent of reading disability will vary with each case. Some patients are able to recognize individual letters but cannot read words (word blindness or verbal alexia), while others can recognize a limited number of words better than letters (letter blindness or literal alexia). Global alexia may be used to describe those who are unable to recognize either.

Other clinical findings commonly associated with alexia without agraphia include visual field defects and defective color recognition/identification. The visual field defects almost always involve the right side and may be quadrantanopic or hemianopic in nature. The acquired defects in color recognition include achromatopsia and color agnosia.

As described by Dejerine in 1892, lesions involving both the left occipital lobe, or its optic radiations, and the splenium of the corpus callosum are responsible for producing alexia without agraphia. Under these conditions, the severely impaired left visual cortex is unable to contribute to visual function and the normal right visual cortex is disconnected from the dominant visual-verbal association center (left angular gyrus) because of the damaged splenial fibers of the corpus callosum. In the pure form, writing and spoken language function is preserved because the left angular gyrus is undisturbed." It is interesting to note that most cases of visual object agnosia and prosopagnosia are associated with alexia without agraphia, however, the converse of this is not true."-"

Stroke is considered to be the most common cause of alexia. Other rare causes include closed head injury, AV malformations, occipital lobectomy, and Alzheimer's disease.233J37 A reversible form of alexia has been observed in patients with operable brain tumois."

Although prospects for rehabilitation are limited, it is important to work with these patients to develop compensatory strategies as well as to educate the patient, family and friends about the condition and its limitations. Some patients are able to decipher some materials by saying each letter aloud to spell the words Once the word is spelled aloud, they are able to recognize it and comprehend its meaning." Understandably, this technique is of marginal use with extensive or complex reading materials, but may be useful enough to allow the patient to read the newspaper headlines and to remain independent in shopping and bill paying. Other patients are able to improve letter recognition by tracing the letters with their f*inge*r*s*.*3*4*3*7 This may be used in conjunction with the auditory word recognition technique just described. A typoscope may be used to help control attention and eye movement, and large print materials are recommended. It also may help to place the reading materials in the patient's good hemifield if a visual field defect is present.

As with many other aspects of rehabilitation, education of the patient, family and friends is of paramount importance. Since the primary symptom is visual, it is often believed that a new pair of glasses will alleviate the difficulties. It is important to dispell this myth and to explain to the patient and his family the reason for the inability to read. In addition, one should consider the severe impact of alexia on the patient's activities of daily living. Reading the newspaper is impossible, difficulties with shopping and bill paying are common, and job loss may occur. Many patients are frustrated and depressed because of the multitude of failures in these areas. It is important to develop activities with attainable goals to provide positive reinforcement and hope for the patient." And finally, it is important to recognize that the patient may feel deeply ashamed of his or her "functional illiteracy."

Ballot's syndrome

Bilateral damage to the parieto-occipital regions may result in a triad of optic ataxia, oculomotor apraxia (psychic paralysis of gaze) and spatial inattention (simultanagnosia).•" First described by Balint in 1907, this syndrome most commonly occurs as a result of stroke, but also may result from tumor, gunshot wounds, Creutzfeldt-Jakob disease, diffuse cerebral anoxia (after open heart surgery), Aizheimer's disease and subacute HIV encephalitis. Partial or complete homonymous hemianopsia is commonly associated." A fourth component of the syndrome was described by Holmes as defective estimation of distance (Balint-Holmes' syndrome).q It is important to realize that each of the four components may occur together or in isolation, thereby producing either complete, or *forme fruste* Balint's syndrome.'7

Optic ataxia

Patients with optic ataxia are unable to visually guide their hand toward an object. They reach for the object in an uncertain manner, demonstrating gross overreaching (pastpointing) or underreaching movements. This may occur despite normal motor and proprioceptive function. In general, unilateral parietoccipital lesions produce optic ataxia involving the contralateral field and band; bilateral defects affect both hands and hemifields"·~ In milder cases, the misreaching involves items in the peripheral field, however, in the most severe form, misreaching occurs while visually fixating the target. This severe form has been described only in association with bilateral lesions in the Balint-Holmes' syndrome.

Prior to testing for optic ataxia, one should document normal motor and proprioreceptive function by having the patient touch his index finger of one hand to the index finder of the other hand. Optic ataxia may then be tested by asking the patient to fixate the examiner's nose as he touches the examiner's finger held in the periphery of the patient's field. The examiner should then hold his finger in front of the patient (avoiding areas of hemifield defect if present) and ask the patient to fixate the finger while attempting to touch it. Alternatively, one may draw two dots or Xs, separated by several inches, on a piece of paper, and ask the patient to draw a line connecting the two targets. The performance is often improved if the patient's finder is placed on the second mark, thus serving as proprioceptive guidance for his or her other hand's movement.

Oculomotor gaxe apraxia

Oculomotor gaze apraxia, or "psychic paralysis of gaze," is the inability to execute purposeful eye movements. As a result, patients have difficulty directing their gaze toward a specific target and once fixation is achieved, it may be easily lost or they may have difficulty in shifting their gaze to a new visual stimulus. This defect in saccades may also occur in response to auditory and tactile-proprioceptive stimuli. Other visuornotor disturbances associated with gaze apraxia include decreased or absent pursuit and the inability to converge in response to an approaching target." Less commonly reported is a fading of the visual percept which is thought to be due to either unstable fixation or rapid visuosensory adaptation. Reappearance of the visual image has been reported to occur with intentional blinking in one patient.

Testing for gaze apraxia should include visual stimuli ("look at my light/hand"), auditory stimuli ("look to where you hear the bell"), tactile-proprioceptive stimuli ("look where I touch your arm"), and verbal commands ("look right," etc.) Pursuit and convergence should be evaluated using visual targets with and without proprioceptive feedback (i.e., patient's finger passively moved by examiner) as well as standard targets. The examiner should test the patient's ability to fixate and maintain steady fixation on objects with and without proprioceptive feedback. And finally, the examiner should evaluate the patient's ability to shift fixation by introducing new visual stimuli in the periphery of the field.

Visual inattention or simultanagnosia

The third component of Balint's syndrome is visual inattention. Despite full or nearly full visual fields, the patients function as though they are looking through a peep hole. This constriction of spatial attention may be so pronounced that the patient is only aware of the single object on which they are fixating." They will fail to perceive other objects outside of their foveal vision. Simultanagnosia is a closely related term and is used interchangeably by some authors to define the third component of Balint's syndrome. This disorder is one in which affected patients are unable to simultaneously perceive more than one stimulus or more than one part of a visual stimulus. As a result, they are unable to recognize the "whole" of a picture despite accurate recognition of its parts. For example, when shown a line drawing of a baseball, a patient with simultanagnosia may become fixated on the seams and consequently mis-identify the object as a railroad track. Another patient with Balint's syndrome recognized the wheel spokes in Figure 5, but mistakenly identified the object as a bicycle. Faulty visual attention and impaired fixation underlie this inability to integrate the parts of a visual stimulus into a whole.

Accurate determination of visual acuities and visual fields may pose a challenge, particularly if the doctor is unaware of the diagnosis. With larger letters, the whole of the letter may fall outside of the foveal vision, and smaller letters may be hard to find due to erratic fixation. It may be of value to test near point acuity, allowing the patient to use his fingers to enhance localization via proprioreceptive feedback. Visual fields may initially appear contracted, but with perseverance and slower movement of test stimuli, accurate results can be obtained.

The examiner may easily screen for the presence of visual inattention or simultanagnosia simply by having the patient describe and identify line drawings and/or pictures in magazines. Formal testing involves the use of complex line drawings with and without overlapping figures."

Rehabilitation

Balint's syndrome can be extremely disabling. It impacts severely on many activities, especially those requiring scanning skills, such as reading, driving and independent navigation. In many respects, these patients are functionally blind.*'

Rehabilitation should include patient and family education regarding the nature of the condition and its functional limitations. The patient may benefit from training in the use of non-visual devices (talking books) and non-visual techniques for activities of daily living. Other compensatory techniques are also of benefit. For example, one patient has reported enhanced television viewing by sitting farther away from the TV to allow more of the picture to fall onto his fovea at any given time. Another patient reports that be is able to minimize effects of optic ataxia by asking people to lay objects on a table, rather than banding them directly to him. The same patient indents the side of soda pop cans so that the opening is always in the correct place for drinking each time be picks up the can.*

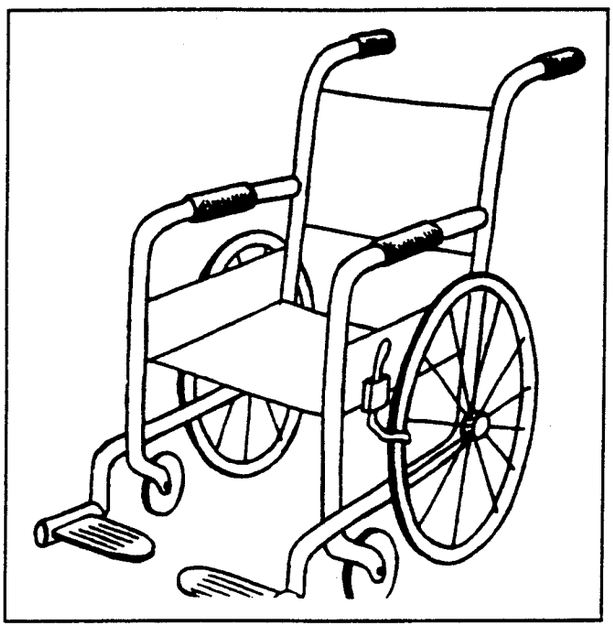

Figure 5: Example of line drawing useful in the treatmentt of simnitanagnocia. (Drawing reproduced with pennurionfrom Kaplan E, GoodglaM H, and Weintraub S. The Bocton Naming Test. Philadelphia: Lea and Febiger, 1983.)

Various techniques aimed at the rehabilitation of visuornotor skills **are** described by Bouska and co-workers' Exercises in "organized visuospatial exploration" for patients with oculomotor gaze apraxia involve the use of a scanning sheet composed of printed rows of letters or numbers. The patient is then asked to circle a specific number or letter each time it appears on the sheet. By requiring the patient to underline the symbols in a continuous fashion as he progresses down each line, the tactile-proprioceptive feedback may stabilize the eye movements. Patients with optic ataxia and impaired estimation of distances may benefit from gross motor spatial training involving non-visual localization of tactile and auditory stimuli. Internal spatial visualization also may be a useful treatment strategy. The training goal with simultanagnosia is to increase the patient's awareness of his peripheral field. As the patient fixates centrally, objects are introduced into the periphery. Progressively smaller and multiple targets increase the complexity of the task.

One should keep in mind that "successful training" at these tasks does not guarantee successful transfer of skills to other activities. In addition, with Balint's syndrome, the interrelation of the three components may preclude the useful application of these techniques.

Table 3: Compensatory strategies in the rehabilitation of patients with visual agnosias

Type of agnosia or functional deficit	compensatory technique
1) visual object agnosia	1) augment visual with tactile/auditory stimuli when possible
2) prosopagnosia involving faces	2A) attempt to identify person by non-facial clues (hair, glasses, etc), voice, clothing, and environment 2B) instruct family and hospital staff to identify themselves and to initiate conversation
3) prosopagnosia involving clothing	3) label all clothing
4) prosopagnosia involving automobiles	4A) memorize car location 4B) memorize license plate number
5) topographical disorientation	5A) use colored dots to mark routes traveled often 5B) utilize verbal instructions for travel 5C) emphasize safety issues (ie, do not drive or travel alone)

Tabte 4: Visual disturbances of higher cortical

Condition	Description	Location of Lesion
visual neglect	failure to attend to objects in affected hemispace	right parietal lobe
visual agnosia	inability to recognize visually presented objects	bilateral occipitotemporal lobes
prosopagnosia	inability to recognize familiar faces	bilateral occipitotemporal lobes
achromatopsia	inability to distinguish colors or hues	inferior occipital lobe, occipitotemporal lobe
environmental agnosia	inability to recognize familiar environments	right occipitotemporal lobes
alexia without agraphia	inability to read despite ability to write	occipital lobe and optic radiations or splenium
Balint's syndrome:		
optic ataxia	inability to visually guide limbs	bilateral parietooccipital lobes
optic apraxia	inability to execute purposeful eye movements	
simultanagnosia	inability to perceive entire picture or to integrate its parts	

Summary

Marks and DeVito emphasize the importance of recognizing these defects in the rehabilitation setting." They describe a patient with visual-spatial disturbances who appeared confused and disoriented, giving the impression of a person with generalized intellectual loss. If the visual disturbances had not been identified, the patient may have been dismissed as a poor candidate for any type of post-stroke rehabilitation, on the basis of confusion or dementia. Similar problems arise in the vision rehabilitation settings. Patients may fail to respond to magnification as expected, and are dismissed as "unmotivated" or "unable to eccentrically view." Accurate diagnosis of the visual disturbance allows the practitioner to redirect rehabilitation techniques to include patient and family education, non-visual compensatory techniques and other techniques as described in this paper. Although a cure can not be effected, the patient is often relieved to find that there is an explanation for bis or her functional difficulties.

In summary, the optometrist is in a good position to play a significant role in the diagnosis and management of patients with visual disorders of higher cortical function. Because of the visual nature of the deficits, the optometrist maybe consulted by the patient, family or hospital staff for evaluation. If the optometrist is knowledgeable in this area, careful history and routine ophthalmic evaluation is often sufficient to alert one to the presence of this type of defect. Table 5 outlines a simple battery of screening tests that can be used to augment the history and ophthalmic eUM,'7 If these acquired disorders of visual perception are suspected, the patient should be referred to a neuropsychologist, neurologist or neuro-ophthaJmologiat for further evaluation. The optometrist may elect to continue his involvement with the patient and provide rehabilitative services.

Table 5: Suggested screening battery for visual disorders of higher cortical function

Screening test	Condition
1) read a paragraph from a newspaper or book	alexia, visual neglect, simultanagnosia
2) copy a line drawing	visual neglect, simultanagnosia
3) describe and identify an object or picture	simultanagnosia, object agnosia
4) identify pictures of famous people	prosopagnosia
5) reach for objects in space under visual guidance	optic ataxia
6) name colors	central achromatopsia, color agnosia

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Footnotes

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