Residual vision with awareness in the field contralateral to a partial or complete functional hemispherectomy

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Abstract—Two patients with unilateral disconnection or removal of the entire occipital lobe were tested for residual vision in their blind field. Using image stabilization to eliminate eye motion artifacts, the central portion of each subject's visual field was tested, beginning 1° from fixation and extending outward to 13.5°. A narrow zone of residual vision was identified along the retinal vertical meridian of each patient. The lateral edge of this zone was generally within 3.5° of the vertical meridian, though extended farther outward (but not beyond 6°) at one field location for each subject. In one patient, it was present in both superior and inferior quadrants; in the other, it was confined to the superior quadrant. Within their zones of residual vision, both patients could detect stimuli and perform simple shape discriminations, but could not name complex line drawings. The patients were aware of their vision within this zone. No residual vision, with or without awareness, was found in areas tested outside these zones. Given the complete absence of visual cortex contralateral to the observed residual vision, alternate structures must be mediating these abilities. Copyright © 1996 Elsevier Science Ltd.

Key Words: hemispherectomy; hemianopia; residual vision; retinotectal; naso-temporal.

Introduction

In humans, damage to primary visual cortex produces a profound state of phenomenal blindness in regions of the visual field which map to the lesioned areas [14, 15]. However, such patients sometimes have demonstrated residual visual function within their scotomas, including the ability to detect and localize stimuli [36, 49, 51] and to perform discriminations based on wavelength [45, 46], orientation [50] or form [11, 52]. In addition, interactions have been reported between blind and seeing field stimuli [30, 40, 43]. Destriate monkeys also show residual vision [18, 32]. Since damage to the primary visual pathway is deemed responsible for the loss of phenomenal vision, such residual abilities have been attributed to secondary visual pathways [6, 40, 51].

However, before accepting secondary visual systems as the mediators of residual visual abilities, one must be certain that the primary visual system is no longer functional. In monkeys, small portions of spared striate cortex can produce a virtual absence of observable visual deficits [21]. Given the variability of the lesions responsible for field defects in humans, it is often difficult to be certain all of striate cortex is inoperative. Recent imaging [5] and psychophysical studies [11, 20, 52] with humans support the idea that remnants of spared cortex can account for some residual visual function.

In order to eliminate the possibility that residual visual abilities are mediated by spared striate cortex, it is necessary to use subjects in which striate cortex has been ablated entirely or completely deafferented. Hemispherectomy patients that have undergone complete removal or deafferentation of a whole cerebral hemisphere satisfy such criteria. In such patients, not only are explanations based on spared striate cortex inapplicable, so are possibilities based on lateral geniculate body (LGB) or collicular projections to ipsilesional extrastriate cortex. Such
a group, therefore, is useful in evaluating residual vision mediated by the ipsilesional superior colliculus (SC) or by projections to the remaining hemisphere.

Several studies with such groups have been conducted. Early demonstrations of detection and localization of bright targets was attributed to light scatter artifacts [53]. When controls were employed to eliminate such artifacts, the hypothesis that these residual abilities are due to the retinotectal projection was strengthened [34, 35]. In recent years, coarse pattern discrimination [38], and detection and localization [39] abilities by hemispherectomized patients, have been reported. In these studies, the subjects were aware of the stimuli presented within their blind field. The extent of the reported residual abilities suggests an interaction between the ipsilesional superior colliculus and the remaining cortical hemisphere (the contralesional hemisphere).

The purpose of the present study was to apply methods (i.e. retinal stabilization) that recently produced data in supporting the hypothesis that some residual abilities are mediated by spared striate cortex [11, 52]. This strict control for eye movements was used to investigate residual vision in subjects with a confirmed absence or disconnection of striate cortex. The first experiment involved mapping the residual detection abilities in the central portion of a subject’s visual field with stimuli placed 1° from the vertical meridian to 13.5° laterally. Two additional experiments assessed the character of the vision within the regions demonstrating residual detection.

Methods

Subjects

In 1990, at age 25 years, subject SE underwent removal of a congenital porencephalic cyst to alleviate intractable seizures. Surgery resulted in a right temporo-parieto-occipital lobectomy [Fig. 1(a) and (b)]. Subsequent to surgery, SE is seizure-free and demonstrates a clinical left homonymous hemianopia without macular sparing [Fig. 3(a)]. Neuropsychological testing indicates that he functions in the average range of intelligence (FSIQ 93; VIQ 90; PIQ 99).

In 1983, at age 18 years, JB underwent surgery for removal of a congenital porencephalic cyst in the left hemisphere. Since he had vision in the right superior quadrant preoperatively, only a subtotal hemispherectomy was performed to avoid exacerbating his field defect. Seizures initially halted, began recurring 3 months later. In 1985, the functional hemispherectomy was completed (Fig. 2). Subsequent to surgery, JB is seizure-free and demonstrates a clinical right homonymous hemianopia without macular sparing (Fig. 3b). Neuropsychological testing indicates he functions near the average range of intelligence (FSIQ 88; VIQ 90; PIQ 88).

Equipment

For all experiments, stimuli were generated with a Macintosh II computer and displayed on a Macintosh color monitor. Head movements were limited through the use of a head rest and bite bar. Subject’s eye motions were monitored with a double Purkinje image eyetracker [8] and the stimulus displays were stabilized retinally with a mirror deflector system [7] attached to the eyetracker. We will refer to this deflector system as the ‘stabilizer’. The resolution of this eyetracker is 1 arcmin [8], and the response time is < 2 msec [7]. This ability to stabilize stimuli retinally makes extended and repetitive stimulus presentations to known retinal positions possible. Subjects viewed the Macintosh display monocularly with their right eye through the stabilizer lens at an effective viewing distance of 57 cm. Other equipment included an IBM PC-AT and a Hewlett Packard 1310 (HP) fast phosphor (p15) X-Y monitor. The IBM was interfaced with the HP monitor and eyetracker via a Data Translation digital to analog (DA) board. To calibrate stabilization, we used a beam splitter to superimpose the image of a non-stabilized HP screen on the image of the stabilized screen. A matrix of five fixation points and a 1° outline square were presented on the non-stabilized screen. The eyetracker offset and gain settings were adjusted until the square surrounded each fixation point as the subject looked to that point. Finally, the stabilizer offsets and gains were adjusted so a 1° circle on the stabilized screen appeared to superimpose exactly on the square and remained so when the subjects moved their eyes. This calibration was checked periodically during each testing session.

Forced choice testing methods were employed. Testing was conducted in a darkened room in order to maximize the likelihood of stimulus detection and discrimination within a field defect [1]. Detection stimuli were flashed several times during each presentation, since patients with visual cortex damage sometimes report the ability to detect flashed or moving objects, but not stationary objects [41]. Unless otherwise noted, black stimuli (< 1 cd/m²) were presented on a gray (10 cd/m²) background in order to reduce the light scatter artifacts which often are associated with bright stimuli on a dark background [4]. Luminance measurements were obtained with a Spectra PR1500 spotmeter.

Stabilized field mapping

Initially, dense stabilized mapping of the blind field for each subject was conducted to explore the boundaries of the field defect and to probe for areas of residual detection. Interval two alternative forced choice methodology was employed to reduce possible criterion effects [29]. On each trial, audible tones defined two successive 600 msec intervals. During one of these intervals, a dark (< 1 cd/m²) stimulus was flashed three times (96 msec on, 96 msec off) on a gray (10 cd/m²) background. At the end of the second interval, the subject was signaled by a tone to indicate which interval contained the stimulus presentation. They indicated their choice with a keyboard press.

Thirty-six contiguous locations were tested within their field defect. Stimuli used were 2.5° square. The area tested began 1° lateral to the vertical meridian and extended 13.5° into the field; vertical extents of the grid reached 10° above and below the horizontal meridian. Four locations were tested within each subject’s unimpaired field, with the medial edge 1° from the vertical meridian and positioned on and 5° above and below the horizontal meridian. Both the subjects were aware that we were investigating the possibility of residual vision within their field defect, so the majority of the stimulus presentations would be within that defect. Subjects were instructed to respond on every trial, guessing the stimulus interval if necessary. We explained that visual information might be processed unconsciously to them and influence their guesses.
Fig. 1. Magnetic resonance axial (a) and sagittal (b) images demonstrating surgical ablation in SE. In 1990, at age 25 years, SE underwent removal of a congenital porencephalic cyst occupying portions of the right temporal, parietal and occipital lobes. Surgery resulted in a right temporo-parieto-occipital lobectomy. Subsequent to surgery, SE is seizure-free. (The remaining left hemisphere is shown on the left-hand side.)

Fig. 2. Computed tomography axial image demonstrating surgical ablation in JB. The frontal and occipital poles were left in place to prevent superficial hemosiderosis and reduce hydrocephalus [48], but they are totally disconnected from the rest of the brain. (The remaining right hemisphere is shown on the right-hand side.)

**Stimulus discrimination**

Pairs of stimuli were presented simultaneously at mirror symmetric locations in each visual field. Stimuli were displayed continuously during a 300 msec interval delimited by tones. The subjects reported if the stimuli were the same or different. In separate sets, simple and complex stimuli were presented. The simple set consisted of a black 2° square and diamond (the square rotated 90°); the complex set consisted of 78 line drawings from a well-studied picture set [44]. The complex figures were chosen with consideration to size, such that none subtended more than 5° of visual angle. Stimuli were presented, centered at one of four different paired locations. For the simple stimuli, these locations were centered at: upfar (+4.75°, +3.75°), upclose (+2.5°, +3.75°), downfar (+4.75°, -3.75°) and downclose (+2.5°, -3.75°); for the complex stimuli, these locations were centered at: upfar (+7°, +3.6°), upclose (+3.6°, +3.6°), downfar (+7°, -3.6°) and downclose (+3.6°, -3.6°).

With the simple stimuli, the diamond was slightly closer to the vertical midline than the square (since the width of the diamond was 2.82°, rather than 2°). However, these stimuli were presented centered 2.5° into the visual field so the medial edge of the stimulus was always separated from the vertical meridian by at least 1°.

**Stimulus identification**

On each trial, a single simple or complex stimulus (from the discrimination experiment) was presented. Stimuli were displayed continuously during a 300 msec interval delimited by tones. The subject's task was to name the stimulus. Responses were typed into the computer by an experimenter for later
Fig. 3. Automatic perimetry demonstrating field defects in each subject subsequent to surgery. Only right eye fields are shown, since our testing was performed with the right eye. (a) Humphrey full field 246 point screening perimetry of ±60° of SE's central visual field. Filled squares indicate blind regions. (b) Humphrey central 30-2 threshold perimetry of ±30° of JB's central visual field. Dark areas indicate blind regions; stippling indicates areas of seeing.

analysis. Simple stimuli were presented at four different locations within each subject's hemianopic field, centered horizontally (negative horizontal values are for JB, positive values for SE) at ±4.75° ('far' stimuli) and ±2.5° ('close' stimuli) and vertically at +3.75° ('up' stimuli) and -3.75° ('down' stimuli). The 'close stimuli' were primarily within areas of spared vision (established by prior detection testing), the 'far stimuli' were outside these areas of sparing. The complex stimuli also were presented both within and outside these areas of residual vision. Only 'close' locations were used to present complex stimuli to JB, both centered horizontally 3.6° to the right of the vertical meridian with one 3.6° above and the other 3.6° below the horizontal meridian. Four locations within the hemianopic field were used to present complex stimuli to JB, two consisted of
the locations used for SE, and two were centered horizontally $-7^\circ$ and $\pm 3.6^\circ$ vertically into his hemianopic visual field. Control locations within each subject's seeing field were used: centered horizontally at $\pm 2.5^\circ$ and vertically $+3.75^\circ$ and $-3.75^\circ$.

**Results**

Stabilized field mapping identified an area in each subject's hemianopic field within which stimulus detection was possible (see Fig. 4). This area forms a band along the vertical meridian generally not wider than $3.5^\circ$, but extending farther outward (but not beyond $6^\circ$) at one field location for each subject. With SE, the band was above and below the horizontal meridian, but only above that meridian with JB. The subjects were aware of their residual vision. During the detection experiments, mean confidence values in areas with sparing were $\geq 3.5$ and $\geq 4.7$ for SE and JB respectively, in contrast to values of $\leq 1.3$ and $\leq 1.2$ in areas without sparing.

Both subjects readily discriminated the simple stimuli. Results are presented in Table 1. With square and diamond figures, SE was correct on 72 out of 80 trials (90%) and JB was correct on 58 out of 60 trials (97%). They were poorer at discriminating complex stimuli, but continued to perform above chance. With these drawings, JB was correct on 20 out of 26 trials (77%, $P<0.01$). Subject SE was at chance above the horizontal meridian, but correct on 65 out of 104 trials below that meridian (63%, $P<0.02$).

Both subjects reliably demonstrated the ability to verbally identify squares and diamonds presented within the zone of sparing. Results are presented in Table 2. Subject SE was correct on 87 out of 96 trials (91%); JB was correct on 20 out of 20 trials (100%). However, neither could identify similarly presented complex figures. In both the discrimination and identification tasks, the subjects performed at chance when stimuli were outside the region with spared detection. Stimuli were always identified correctly in each subject's seeing field.

**Discussion**

Since residual vision was limited to a region proximal to the vertical meridian, we were concerned about the possibility of eccentric fixation. If our subjects fixated eccentrically (i.e. looked 'off to the side' when we instructed them to look 'at' the fixation point) during the calibration of the eyetracker, stimuli presented to their blind field could have fallen within their seeing field. It is unlikely that eccentric fixation can account for the irregular distribution of residual vision along the lateral edge of the zone of sparing demonstrated by both subjects. We nevertheless obtained acuity profiles across the subjects' point of fixation using a Tübinger perimeter [19]. Both subjects showed an acuity peak at the fixation point, with acuity becoming non-measurable $1^\circ$ into their hemianopic field, a pattern indicative of foveal fixation that is in character with the other crude discrimination and identification abilities of these subjects. Huber [16, 17] demonstrated similar limited sparing in the contralateral hemifield using a Tübinger perimeter and manual stimulus positioning.

Although we used dark stimuli and provided a buffer zone between our stimuli and the vertical meridian, the possibility of light scatter artifacts could not be totally eliminated. Recent studies have concluded that light scatter, both within the environment [22] and due to the reflective properties of the eye [9], can account for many demonstrations of residual vision. However, as with eccentric fixation, this potential artifact cannot account for the uneven spatial distribution of the residual vision in our subjects.

Artifacts due to subjects' eye motions were essentially eliminated by our use of retinal stabilization. The resolution and response time of the eyetracker and stabilizer were sufficient to maintain stimuli in the hemianopic field of our subjects even in the face of saccadic eye motions of several degrees.

We have therefore demonstrated residual vision in the
Fig. 4. Schematic representations of stabilized visual field detection results for SE (a) and JB (b). Open squares represent above chance performance, filled squares indicate chance performance. Values presented at each location are percent of correct detections, number of trials run (n) and significant P-values.
blind field that is not likely due to eccentric fixation, light scatter or eye movements. The fact that the subjects were aware of their residual vision contrasts with more reports of blindsight, a term used to describe residual vision in the absence of acknowledged awareness [51]. In this regard, our data are similar to those previously reported by Ptito and colleagues [38, 39] with a different group of hemispherectomy patients. However, these other patients demonstrated residual detection and localization abilities in their blind field at greater eccentricities than those reported here. The reason for this discrepancy is not clear, but individual differences are evident in the previous group of patients and their residual abilities increased with proximity to the vertical meridian [39].

What is the source of our subjects' residual vision? One possibility, which has been invoked to explain other cases of residual vision following hemispherectomy [34, 35, 38, 39], is that these abilities are subserved by a direct retinal projection to the SC. Such a pathway could be mediating SE and JB's residual vision, possibly via an intertectal connection to the remaining hemisphere. Recent quantitative cytoarchitecture and cytochrome oxidase activity analyses of the effects of early hemispherectomy in the vervet monkey further support this notion, showing that the ipsilesional SC is affected by the loss of cortical input but remains a functional subcortical endstation for analysis of visual information. Additionally, cellular integrity and metabolism of the SC are much less affected than those of the dorsal LGB [2, 37]. Similar residual SC function could occur in human subjects with pathologies present at an early age, such as SE and JB. Such an explanation, however, does not fully account for the residual visual abilities we found.

An alternative explanation is that these abilities are subserved by a zone of naso-temporal overlap. Most psychophysical studies with humans [10, 31, 47] and anatomical studies with monkeys [3, 13, 24] suggest any zone of naso-temporal overlap is narrower than the band of sparing we found in our subjects (though, see Ref. 12). In the absence of a normal pathway during development, the zone containing uncrossed retinal projections from the nasal hemiretina and crossed projections from the temporal hemiretina may have expanded. This would provide the intact hemisphere with increased access to the ipsilateral visual field. Recent developmental studies in the cat and monkey are in accord with this possibility. In the cat abnormal development following a unilateral optic tract section results in a significant increase in the number and size of contralaterally projecting alpha and beta (but not gamma) cells within the temporal retina [25, 26, 42]. Such developments tend to favor the central visual field [33]. In the neonatal monkey the normally sharp naso-temporal division [23, 28] begins to break down during abnormal development caused by a retinal lesion [27]. Although the porencephalic cyst was not evacuated until JB was 18 and SE was 25 years of age the pathology in these subjects was present at birth. Developing retinofugal projections lacking their normal cortical projection area could have been re-routed to the intact hemisphere in these subjects.

The character of residual vision demonstrated by our subjects is commensurate with this possibility. While limited, their abilities included crude form discrimination with occasional complex form discrimination. Such abilities normally require cortical processing. Given the complete lack of ipsilesional striate and extrastriate cortex, any cortical processing must have occurred in their intact hemisphere. Expansion of the zone of naso-temporal overlap is a likely candidate for enabling access to that hemisphere, although reorganization of the retinotectal pathway may also contribute. Additional testing of other residual abilities (such as the localization of stimuli with eye movements) may further define the characteristics of this residual vision and serve to dissociate SC and cortical processing. This, in turn, may help to further clarify the anatomical pathway mediating such abilities.

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