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Comparison of three Ways to Assess Residual Vision after Macular Vision Loss

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Abstract: This research addresses the problems faced by patients with macular vision loss who are trying to adopt new viewing strategies. The hypothesis is that these problems are tightly connected with the detailed topography of residual vision in both eyes. In order to assess this topography in a quick and cost-effective way, a computerized test was developed and is here compared to already established methods. The results show that existing methods of analyzing visual field properties do not yield the information necessary to guide patients with macular vision loss through the difficult period of adopting new viewing habits. In contrast, our test provides the necessary data and does so in much less time and with less fatigue for the patient.

Introduction

Patients with maculopathies have lost the ability to perform tasks requiring high spatial resolution. Our work focuses on patients with a dense (or "absolute") scotoma including the fovea and parts of the macula. The most typical patients are those suffering from age-related maculopathy (ARM). The patients' experience is that foveated visual objects disappear. The majority of them complain most about the loss of the ability to read, while their mobility often remains quite good.

Reading requires, among others, two things: Adequate spatial resolution and the ability to make accurate goal-directed eye movements. Spatial resolution (visual acuity) is non-linearly distributed across the visual field [1, 2, 3], a consequence of anatomy that cannot be changed (e.g. cone density, ganglion cell density, ratio of convergence of cone outputs onto ganglion cells etc.). Consequently, if the fovea is damaged and a perifoveal area has to be used instead, adequate magnification has to be provided to compensate for the lack of spatial resolution. In practice, however, this will not be enough to compensate for the foveal damage. This is demonstrated by the fact that patients with newly diagnosed ARM have great difficulties reading.

In order to understand these additional difficulties, we have to look at the other necessary element, i.e. eye movement control. How does the visual/oculomotor system know where to look next? The involved mechanisms are poorly understood, but we would like to venture a hypothesis. Imagine the following experiment: A normally sighted subject looks at the center of an array of 20 degrees diameter showing a large number of targets for only 100 msec. When asked whether the array contained one target that looked different from all the others, the subject will not be able to answer the question. Even if the size of the targets were scaled to compensate for the declining resolution in the periphery, the result would be the same.

However, if the subject received a spatial cue shortly before the appearance of the array indicating the location to be judged, perception of the target at that location would be clear and the answer would be easy. The reason is that, in this case, the subject will have an expectation as to where the critical target will appear and, thus, will be able to shift attention to the cued location. A previous research project in our laboratory revealed that there are two components to focal visual attention: A sustained one that is subject to volition and a transient one that acts more reflex-like and only up to 500 msec after target appearance [4]. Both can enhance perception and both are important for patients with macular vision loss. It is obvious for the sustained component, because it can be shifted at will to any location in the visual field. This is exactly what ARM patients need, as the nature of the task they have to solve is: "Look at one thing and attend to another".

Furthermore, in regard to transient focal attention there is evidence from a variety of different research approaches indicating that it can lead goal-directed eye movements [5, 6]. To provide a theoretical background for the present report, we hypothesize that during reading, the two components of focal attention may play the following roles in reading. The sustained component defines the resting point (normally the fovea), while the transient component jumps ahead by a few letters (in this text to the right) and is re-united with the fovea by the ensuing fast eye movement, then jumps ahead again etc. One can visualize the interaction between transient focal attention and the fovea as a caterpillar-like form of locomotion. In other words, during reading the attentional focus wanders back and forth in a horizontal streak around or to the right of the fovea. Consequently, the ARM patient who wants to re-learn how to read with any degree of fluency has to learn how to let the attentional focus perform the same movements around an intact retinal spot outside the damaged area. Fluency cannot be achieved by reading letter-by-letter, the horizontal extent of this spot should be enough to accomodate 4-6 letters [7, 8]. This is the reason why ARM patients with a small island of intact vision near the fovea may have good letter acuity, but will still be unable to read at an acceptable speed.

The demand on letter size grows even faster than explained by the decline of resolution (or letter acuity) with eccentricity [7], probably because the retinal periphery is more susceptible to lateral interactions between letters, i.e. "crowding" [9, 10]. Hence, the required retinal area has to be big enough to accomodate 4-6 letters of sufficient size. Visual attention, in spite of its high mobility, always returns to the fovea (Nakayama and Mackeben., 1989; Mackeben and Nakayama, 1993). This aspect also connects it to the center of the coordinate system required for goal-directed eye movements as well as for precise localization of objects in the visual world. It has been shown that, with time, patients with macular vision loss develop not only a "preferred retinal locus" (PRL), but also a new center of the retina [11]. Having a PRL in the periphery will obviously sacrifice resolution, which can be compensated for by optical means, i.e. by the use of magnifying glasses. Developing a new center of the oculomotor control system, however, requires a process of re-learning that, for some unknown reason, is harder for some patients than for others.

How do ARM patients deal with this challenge? With time, a certain percentage of them learn to practice "eccentric viewing" on their own, maybe as many as 25% [12]. Because of the fact that this learning process happens without outside assistance, the process is often referred to as "spontaneous". This should not imply speed or lack of effort, because some patients report that the process can take years and requires a great amount of effort of will and practice. During this process, the patient may develop one PRL for one task and another one for a different task.

On the other hand, some rehabilitation specialists think that among the patients who do not discover eccentric viewing on their own, there is a small percentage who cannot be helped even by the most sophisticated instructions or training procedure guided by an eye care specialist. Considering the age of ARM patients, it is likely that a combination with other diseases and impairments contribute to this condition. However, it should also be mentioned that the existence of such a group is denied by some practitioners of eccentric viewing training. According to the extensive study conducted by Nilsson and Nilsson in Sweden [12], the reason may be that short-term effects can be seen in all patients, while only 80% showed substantial long-term benefits after an average of 5 years. Moreover, the failure to show such effects in half of the remaining 20% could be explained by reasons other than the patients' eye condition.

In trying to understand the differences between individual patients, the obvious variable to look at first is age, since it has been shown that age alone can impose serious limitations on a person's ability to perform visual tasks [13, 14]. However, clinical experience in the Low Vision Service headed by one of us (AC) shows that age is a poor predictor for this purpose. Next would be visual acuity for letters, which may reflect the size of the central scotoma. However, it is equally unable to predict a patient's chance to discover a successful new gaze strategy.

Even if two extreme groups were clearly definable entities, it would leave a large group of patients who do not belong to either of them. The majority of ARM patients can be expected to be over 60 years of age. Under these circumstances, time is a precious commodity, and to have to wait for years for an improvement that may or may not happen "spontaneously" is unacceptable.

Our work has two goals: To understand the mechanisms underlying the differences between individual patients, and to address the needs of the patients who are between the extremes outlined above. These are the patients who do respond to instructions and who can be trained to use eccentric viewing to overcome the effects of macular disease, but for some reason never discover it on their own [12].

We believe that the eye care specialist should be able to give explicit instructions based on the individual patient's condition. Our working hypothesis is that this can happen only through a careful assessment of the detailed topography of residual vision. It should be noted that this approach is not the same as mapping scotomata, but rather concentrates on the areas with intact vision, particularly the ones of potential value for eccentric viewing.

Methods

Three Tests: We have looked at different methods of assessing residual vision and selected two of them for a comparison with a procedure that we have developed in our laboratory [15, 16]. First, there is the clinically well-established Humphrey Visual Field Analyzer. As we are most interested in the center of the visual field, we use only Humphrey's "10 - 2" routine, which tests the central 20 degrees. The second is the completely computerized method used by the Ophthimus test, using only their "Centring" routine, again analyzing only the central 20 degrees. Our own mapping test is described in more detail elsewhere [16]. It is also implemented on a small computer, but shows some significant differences from the other two because of the fact that it has been designed specifically for the purposes of the research objectives stated above. A summary of the features of the three different tests is shown in Figure 1.

Figure 1: Comparison table showing the most important features of the three tests.
For the purpose of this discussion, we would like to mention only a few cardinal points. All three tests use tachistoscopic stimulation, while the targets differ greatly: The Humphrey uses small light spots on a dark background, the Ophthimus dark double-contour rings on a light background, and our own test uses dark letters and groups of letters on a light background.
Both, the Humphrey and the Ophthimus test determine detection thresholds. In contrast, our test measures discrimination thresholds for letters and words. This is relevant because reading requires letter/word recognition, and we want our results to relate directly to the ability to read. There is a rich body of psychophysical literature showing that discrimination thresholds are higher than detection thresholds [18]. Detection alone, though necessary, will not do the patient any good, and knowing its threshold will not allow any meaningful conclusions regarding the usefulness of a retinal location for reading. Hence, detection of light spots as used in the Humphrey test may be sufficient to map a scotoma, but not enough to indicate the usefulness of a retinal locus for reading. Because of other incongruencies, we cannot confirm yet whether the claim that the detection thresholds for high spatial frequency-filtered rings used in the Ophthimus are indeed equivalent to discrimination thresholds [19].
There are also differences in how the thresholds are determined: The Humphrey varies contrast via brightness of the light spots, the Ophthimus varies contrast and size of the rings, and our own test normally varies only letter size (five different contrast levels are available for special investigations).
The second important difference is that the Humphrey and Ophthimus procedures measure thresholds in all available test spots, while our test first reduces the number of test points, so that valuable time is saved and unnecessary strain on the patient is prevented. The selection of test spots uses the criterion of what might be helpful for serving as a preferred retinal locus for reading. For instance, a tiny intact spot relatively close to the fovea may render good letter acuity, but is likely to be useless for reading, because it allows reading only letter-by-letter and requires that the patient repeatedly performs saccades into a scotomatous area. Investigations using scanning laser ophthalmoscopes have shown that in selecting a preferred retinal locus (PRL) patients with maculopathies tend to sacrifice spatial resolution (i.e. acuity) for a larger area even if it is farther away from the fovea [20, 21].
A third point of great importance is the reliability of the topographic data, which hinges on the issue of gaze stabilization. It should be kept in mind that patients without macular vision cannot "fixate" in the usual sense, so that a central fixation mark is useless. The Humphrey test's use of four little light spots for gaze stabilization makes it very hard for patients to hold their eye still, because any or all lights might fall into the scotoma. The other two tests use the same principle, i.e. peripheral landmarks that cover a larger area (spokes pointing at the center of the screen). Quantitative data confirming the validity of gaze stabilization with peripheral landmarks have been reported before [15,17].
Patients: For this pilot study, we tested three ARM patients with very different visual acuity ratings (20/300, 20/120 and 20/40) and ages between 74 and 79 using all three tests.

The measurements for any one patient using all three methods were conducted on the same day in order to avoid day-to-day variations of performance. The sequence of the tests was different for each patient (as indicated by the ordinal numbers in Fig. 3).
The procedure of our own test consists of two phases, i.e. a first run of 36 trials in pseudo-random sequence (4 for the center and one each for 32 peripheral locations) and the ensuing threshold determination for up to three "candidate" spots that the program identifies as promising for eccentric viewing. For the purposes of this pilot study, we were less interested in absolute thresholds but rather in the accuracy of the topography. At the same time, we had to be very careful not to over-challenge our patients' willingness to cooperate in this very fatiguing series of measurements. We therefore limited our test to the first round of 36 trials. Normally, the determination of the thresholds can add 1 - 3 minutes to the overall test duration. It should be kept in mind that the size estimation we perform to determine which target size set to use (see [16] for details) usually get us rather close to the threshold anyway (see below).

Results

Figure 2 shows the results of the Humphrey, Ophthimus and our own tests, respectively, for one patient. The obvious difference between the Humphrey and our test is the fact that the scotoma is not only shaped differently, but also in a different location, which was most likely caused by instability of gaze in the Humphrey test ("fixation" lights lay within the scotoma). For unknown reasons, the Ophthimus chart does not show the eccentric location of the macular scotoma seen in the results of our test, where the usable part of the retina is clearly located in the left visual field. This finding was verified by mapping the scotoma in a computerized, hand-run procedure that allowed the examiner to place the location to be tested anywhere in the circular display area (see [16] for details). Furthermore, the patient confirmed the result of our test with credibility, because she uses eccentric viewing consciously and successfully. We attribute the divergence of the Ophthimus results from those of our own test in part to the general insecurity we felt in interpreting the graphic display of that test.
One of the most conspicuous differences was the time required to take the tests (see Figure 3). The means for the Humphrey, the Ophthimus and our test were 17:16 min, 9:53 min and 3:33 min, respectively. We expect these durations to be proportional to the amount of fatigue experienced by the patients.

Figure 2: Graphic representations of the results of the the Humphrey (upper left), the Ophthimus (upper right) and our own test (bottom). Darker areas in the Humphrey graph denote higher thresholds, as do larger rings in the Ophthimus. The filled circles in the graph from our test show locations where the target was not detected at all, half-filled circles mean that the letter was detected but could not be identified, open circles show locations where the letter was correctly identified. Note the conspicuous differences in the topography of vision loss as well as in the durations of the tests (see also Fig.3).

Figure 3: Comparison of the test durations for all three patients. The encircled ordinal numbers denote in which order of sequence the patient took the test. Each patient took all three tests during one day (within maximally 2 hours).

In regard to the size threshold, we used the rule of thumb described elsewhere [16] and chose a set of 20 to 35 pixels for our test, which left this patient very little "acuity reserve" [8]. Consequently, the bottom of Fig. 2 shows that the letter size of 30 pixels (70 minarc at this distance) used for the two "good" spots on the third ring to the left and lower left of the center is the approximate equivalent of a visual acuity rating the patient received in the doctor's office (20/300).

In addition, two of our patients complained that the Humphrey test was an "unpleasant experience" because of the difficulty in using the diamond formation of four "fixation" lights in an otherwise empty visual environment to stabilize their gaze. This may also account for the considerable differences between the Humphrey and our test concerning scotoma localization in patient KE (see Fig. 2). The match was only slightly better in the second patient. The third patient showed only very subtle deficits, most of them only when we used low contrast letters. She has severe ARM in one eye (v.a. 20/500), while her other eye is still largely unaffected (v.a. 20/40). We have studied only her "good" eye over a period of 16 months in order to detect possible early signs of progression of ARM. The subtle defects we saw here were not picked up by the Humphrey test at all. The Ophthimus test showed agreement only in regard to one location, were the target was not detected. Again, the subtler effects (letter detected, but not recognized) were not picked up. This last fact casts some doubt on the claim that the detection threshold for the ring targets should be the same as a discrimination threshold [19].

In conclusion, we would like to emphasize that this discussion does not intend to diminish the validity of the Humphrey and Ophthimus tests for general field testing. They were, after all, not specifically designed to examine ARM patients from our very specialized point of view. We wish to point out, though, that there is a need for a specialized test like ours that can aid in the assessment of the topography of residual vision for the purpose of rehabilitation of patients with dense macular scotomata.

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