Copyright © Association for Research in Vision and Ophthalmology

Contrast Thresholds for Letter Identification in Retinitis Pigmentosa
Kenneth R. Alexander, Deborah J. Derlacki, and Gerald A. Fishman

To assess mechanisms of foveal vision loss in retinitis pigmentosa (RP), contrast thresholds were measured for the identification of Sloan letters at four adapting field luminances (0.4, 1.4, 2.4, and 3.4 log td) in a group of 16 patients with RP who had best-corrected Snellen visual acuities of 20/30 or better, minimal or no posterior subcapsular cataracts, and no atrophic or cystic-appearing foveal lesions. Letter contrast sensitivities of the patients with RP were reduced below those of a group of ten subjects with normal vision for all letter sizes and at all adapting field luminances. The overall pattern of these results indicated that neither a reduced quanta! absorption by foveal cones nor spatial undersampling from a loss of foveal cones accounted for the reductions in letter contrast sensitivities. The findings were most consistent with a uniform increase in intercone spacing in the foveas of this group of patients with RP and mild visual acuity loss. Invest Ophthalmol Vis Sci 33:1846–1852, 1992

Studies of the visual acuity of older individuals with normal vision and patients with age-related macular degeneration have found that the degree of visual acuity loss observed under standard clinical test conditions of high contrast and relatively high luminance may not predict the degree of foveal impairment under conditions of low contrast and low luminance. Similarly, our recent investigation of the foveal acuity–luminance functions in a group of patients with retinitis pigmentosa (RP) showed that they tended to have a loss of visual acuity at low adapting luminances that was disproportionately greater than the acuity loss at high adapting levels. Whether patients with RP also have a disproportionate loss of acuity at low contrasts has not been established, although evidence suggesting such a relationship has been presented. To address this issue, we examined more extensively the effects of contrast and luminance on foveal spatial vision in patients with RP by measuring their letter contrast sensitivity functions (LCSFs) at various adapting field luminances and over a range of Sloan letter sizes.

Currently, the exact explanation for the foveal acuity loss of patients with RP is uncertain. Proposed hypotheses include a reduced light-catching ability of foveal cones and a decrease in cone photoreceptor spatial density. These hypotheses are consistent with histologic studies of foveal cone photoreceptors in donor eyes from individuals with RP, which have shown shortened and/or disorganized cone outer segments and a reduction in the number of photoreceptors per unit area of the fovea. Our recent study of foveal acuity–luminance functions in a group of patients with RP and mild acuity loss suggested that a decreased quantal absorption by the foveal cones was not the primary explanation for the reduction in visual acuity because acuities were not improved by increased levels of chart illumination. The pattern of visual acuity loss instead was most consistent with a decrease in the spatial density of their foveal cones.

Two distinct ways in which a change in foveal cone spatial density might occur in patients with RP are (1) as a result of a uniform increase in foveal intercone spacing or (2) as a consequence of a random loss of foveal cones. These two possibilities have different consequences for spatial vision. A uniform increase in intercone spacing with preservation of neural connections to the postreceptoral mechanisms or analyzers that are presumed to mediate spatiotemporal vision is equivalent to a change in the spatial scale of the fovea. Such a change in spatial scale would result in a lateral displacement of the LCSFs of patients with RP along a size axis, with a comparable degree of displacement at all adapting field luminances.
nances. Alternatively, a random loss of foveal cones would lead to spatial undersampling in postreceptoral spatial mechanisms, which should have the greatest effect on the highest spatial frequency analyzers.\(^{10}\) The effect of undersampling should be most evident at high adapting luminances, at which high spatial frequency mechanisms are most sensitive,\(^{12}\) with the result that the loss of contrast sensitivity should be greatest for small letters. We analyzed the LCSFs of patients with RP to test these two hypotheses concerning possible underlying mechanisms of foveal visual acuity loss.

**Materials and Methods**

**Subjects**

Sixteen patients (12 men and 4 women) with typical RP or Usher's syndrome participated in the study. Based on previously established genetic criteria,\(^{13,14}\) one patient had autosomal recessive RP, nine had isolated cases of RP (no other family member was known to be affected), three had type 2 Usher's syndrome (a recessively inherited variant of RP accompanied by a congenital neurosensory hearing impairment), and three had RP of uncertain genetic type. These patients with RP had best-corrected pretest Snellen visual acuities ranging from 20/15 to 20/30, minimal or no lens opacities, no atrophic-appearing foveal lesions, and no macular cysts, although seven had epiretinal macular membranes. Their ages ranged from 24–53 yr (median, 32.5 yr). Results obtained from testing these patients were compared with those from ten (6 men and 4 women) age-similar control subjects with normal vision (age range, 23–54 yr; median, 38 yr). All control subjects underwent an ophthalmic examination and had normal-appearing fundi, with best-corrected Snellen visual acuities of at least 20/20. Informed consent was obtained from all participants after the nature of the testing procedures had been explained fully.

**Procedure**

Before testing, the pupil of the tested eye was dilated with phenylephrine hydrochloride 10% and tropicamide 1% drops, and the tested subjects were dark adapted for 15 min. Contrast thresholds for the identification of isolated Sloan letters were measured with a computerized video display system (BVAT IISG, Mentor O & O, Norwell, MA) in a darkened room. Isolated Sloan letters with visual acuity values ranging from 20/15 to 20/300 and of variable contrast were presented in the center of a rectangular background that subtended 2.2° horizontally and 1.6° vertically.

The letters were of negative contrast, with contrast (C) defined by the Weber relationship:

$$C = \frac{(L_T - L_B)}{L_B}$$

where \(L_B\) is the background luminance, \(L_T\) is the letter luminance, and \(L_B\) is less than \(L_B\). Test subjects viewed the video monitor monocularly through a 5.5-mm artificial pupil and an appropriate correction in a phoropter. The test distance was 5.5 m. Display luminance was varied by placing Kodak Wratten No. 96 neutral-density filters (Eastman Kodak, Rochester, NY) of appropriate values in a holder attached to the eyepiece of the phoropter. Retinal luminances of the backgrounds were 0.4, 1.4, 2.4, and 3.4 log td, as derived from calibrations with a Spectra Spot photometer (Kollmorgen, Newburgh, NY).

Letter contrast sensitivities were completed first at the lowest adapting level and then at each of the three higher adapting levels in succession. The tested subjects adapted for 1 min to each background luminance before testing commenced at that level. At each adapting level, the order of the letter sizes was randomized. Then, an initial estimate of threshold contrast was obtained for each letter size with an ascending method of limits. During each trial, a randomly chosen letter of the appropriate size and contrast was presented continuously until either the subject responded or a maximum duration of 10 sec had elapsed. The tested subjects were instructed to identify each letter as it was presented and were encouraged to guess if they were uncertain. The initial contrast was set to a level that was below the estimated threshold, and contrast was increased by one step (0.2 log units) between trials until a letter could be identified correctly. The lowest contrast at which a letter was identified correctly was used as an initial estimate of threshold.

The threshold contrast for letter identification then was measured according to the Pelli-Robson procedure,\(^{15}\) as modified for our testing apparatus. Three isolated letters were presented at each contrast level, beginning at a contrast value that was approximately 0.6 log unit above the initial threshold estimate. If all three letters were read correctly, the contrast was reduced by one step, and another series of three letters was presented. The trial ended when the subject was unable to identify at least two of the three letters correctly at a given contrast level. The threshold contrast for each test condition was defined as the lowest contrast at which two or more letters were identified correctly. Two measurements were made for each letter size at each adapting level, with the letter sizes presented in counterbalanced order. The two contrast threshold values for each letter size were averaged.
Sensitivity was defined as the reciprocal of the threshold contrast.

Results

The mean LCSFs for the normal control subjects at each of the four adapting levels are presented in Figure 1. Letter contrast sensitivity increased with increasing adapting field luminance, a finding that is consistent with studies of contrast sensitivity for sinusoidal gratings. The solid curves in Figure 1 are the least-squares best fits of the log form of an exponential equation that has been used previously to describe contrast sensitivity functions for sinusoidal gratings:

\[ s = a(1/MAR)^p e^{-n(1/MAR)} \]  

(2)

where \( s \) represents sensitivity; MAR is the minimum angle of resolution; \( a \) and \( p \) are scaling parameters representing vertical and horizontal positions, respectively, on log-log coordinates; and \( n \) is the slope of the attenuation at low frequencies. In the current analysis, \( n \) was set to 0 because no low-frequency attenuation was apparent for the range of letter sizes used. This function provides a reasonable fit to the mean LCSFs of the normal control subjects.

Examination of the best-fit parameters \( a \) and \( p \) for the mean normal LCSFs indicated that, as adapting field luminance increased from 0.4–1.4 log td, the primary change was a vertical shift of the LCSF, corresponding to an increase in contrast sensitivity that was equivalent for all letter sizes. As luminance increased further from 1.4–3.4 log td, there were approximately equivalent horizontal and vertical displacements of the normal LCSF. As a result, the greatest increase in contrast sensitivity occurred for the smallest letter sizes over this higher luminance range.

The letter contrast sensitivities of the patients with RP (Fig. 2, symbols) were reduced below those of the control subjects (Fig. 2, solid lines) at each of the four adapting field luminances. As seen with the control subjects, the mean LCSFs of the patients with RP were well-described by Equation 2 (Fig. 2, dashed lines). A comparison of the best-fit values of the parameters \( a \) and \( p \) for the two groups indicated that the curves for patients with RP were shifted downward and to the left of those of the control subjects by comparable amounts at all luminances. This relationship between the LCSFs of patients with RP and those of the control subjects is illustrated further in Figure 3.

The mean data for patients with RP have been translated as a group upward and to the right of the curves in Figure 2 by an amount equal to the mean difference in the values of \( a \) and \( p \) between the two groups (Fig. 3, calibration bars). When the data for the patients with RP were translated in this way, they super-
imposed well on the curves that described the normal data (Fig. 3, solid lines).

The LCSFs can be used to derive the effect of letter contrast on the visual acuities of those with RP (Fig. 4). The two curves in this figure describe the relationship between log MAR and log contrast at the highest adapting level for the control subjects and those with RP. The data and curves have been replotted on transposed axes from Figures 1 and 2. For the normal control subjects, the value of log MAR increased (i.e., visual acuity decreased) as the contrast was reduced, as expected. A similar relationship between log MAR and log contrast was evident for those with RP. However, the curve for the patients lay to the right of that of the control subjects (corresponding to a vertical displacement of the mean LCSF of those with RP, Fig. 3). Consequently, the vertical separation between the two curves was largest at low-contrast levels; that is, the greatest loss of visual acuity for those with RP relative to the control subjects occurred at the lowest contrast levels.

Because the mean LCSFs of the patients with RP could be superimposed on those of the control subjects by a simple transformation (Fig. 3), this indicates there was a normal relationship between light adaptation and letter identification in patients with RP. This relationship is illustrated further in Figures 5 and 6. Figure 5 plots the mean decrement thresholds for letter identification (log |L_D - L_T|) as a function of log background luminance (L_D) for the control subjects. Each curve represents the results for a different letter size. At low adapting levels, the decrement thresholds rose with a slope of approximately one half; the slopes
approached Weber's law (Fig. 5, dashed line) at the higher adapting levels. The slopes of the letter decrement threshold functions were nearly the same for all letter sizes tested, a finding that agrees with a recent study of foveal increment threshold functions for the detection of various sizes of test flash.\textsuperscript{18}

The short horizontal bars in Figure 5 represent the maximum available decrement threshold (ie, \(L_d = 0\)) at each adapting field luminance. For the smaller letter sizes, data points are absent at the lower adapting levels because the decrement thresholds for letter identification exceeded these maximum values. The decrement threshold functions of the patients with RP showed a corresponding pattern of results for the detection of various sizes of test flash.\textsuperscript{18}

A more direct comparison between the decrement threshold functions of the two groups is illustrated in Figure 7 for the largest letter size (20/300). The dashed curve represents the prediction of a reduced quantal absorption by the foveal cones of those with RP; that is, it corresponds to a displacement of the normal decrement threshold curve along a 45° axis by an amount equivalent to the threshold difference between the two groups at the lowest adapting level. By contrast with this quantal catch prediction, the results for the patients with RP lay above the normal results by the same amount at all background luminances.

This finding was confirmed by a repeated-measures analysis of variance on the results for both 20/200 and 20/300 letters, which were the only conditions for which data were available for all subjects tested at all adapting levels. For both letter sizes, there were significant main effects for diagnosis \(F_{3,35} = 71.9\) [20/200] and 70.9 [20/300], \(P < 0.01\) and luminance \(F_{3,35} = 611.8\) [20/200] and 786.1 [20/300], \(P < 0.01\), as expected. However, in neither case was the interaction between diagnosis and luminance statistically significant \(F_{3,35} = 0.06\) [20/200] and 0.11 [20/300], \(P =\) not significant. The nonsignificant interactions indicate that the decrement threshold functions of the patients with RP were displaced vertically from those of the control subjects by a comparable amount at all adapting field luminances.

**Discussion**

Contrast sensitivities for the identification of Sloan letters were reduced below normal at all letter sizes and at all adapting field luminances in this group of patients with RP and mild visual acuity loss. Of particular interest, the LCSFs of these patients were characterized by both horizontal and vertical displacements from the LCSFs of the control subjects, with a comparable degree of displacement at all adaptation levels (Fig. 3). These results have several implications for
assessing the mechanisms of visual acuity loss and characterizing the degree of functional impairment of these patients.

First, the decrement threshold functions for letter identification derived from the LCSFs indicate that, in agreement with our previous study of acuity–luminance functions in patients with RP, a reduced quantal absorption by foveal cones is not the primary explanation for their foveal visual acuity loss. A reduced quantal absorption by the foveal cones would result in a loss of sensitivity to both background and letters, such that the letter decrement threshold functions of these patients would be displaced by 45° from those of the control subjects. Contrary to this expectation, the decrement threshold functions of those with RP were elevated above normal at all adapting field luminances (Fig. 7).

Instead, our results support the hypothesis of a change in the spatial scale of the foveas of patients with RP, a conclusion that agrees with our previous study of acuity–luminance functions in such patients. As already noted, a change in spatial scale could result from either a uniform increase in intercone spacing or a random loss of foveal cones. However, the finding that the LCSFs of those with RP were displaced from normal to a comparable degree at all luminance levels is inconsistent with the hypothesis that changes in letter contrast sensitivity were primarily the result of undersampling from a random loss of foveal cones. Such undersampling would result in a loss of contrast sensitivity predominantly at high luminance levels and at high spatial frequencies (ie, small letter sizes). Rather, these results are most consistent with the hypothesis that there was a uniform increase in intercone spacing in the foveas of our patients with RP. Such an increase in intercone spacing would produce an effective minification of the neural image (micropsia) that would be of a comparable magnitude at all adapting levels.

In addition to a change in spatial scale, our results show that patients with RP had an overall reduction in contrast sensitivity for letter identification, including letters with a visual acuity equivalent as large as 20/300. This finding is consistent with reductions in contrast sensitivity at low spatial frequencies that have been observed in previous studies using sinusoidal gratings in patients with RP. Currently, the explanation for the overall loss of contrast sensitivity is uncertain. One possibility is a decreased contrast of the retinal image resulting from light leakage between foveal cones. For example, patients with RP and mild acuity losses can have a reduced Stiles-Crawford effect, which has been hypothesized to result from an enlargement of cone inner segments. A reduction in the Stiles-Crawford effect would entail both a broadened acceptance angle of incident light and light leakage between foveal cone photoreceptors, which would, in turn, decrease the effective contrast of the retinal image. An enlargement of cone inner segments also could explain the generalized increase in intercone spacing suggested by our results. Whether this is the correct explanation for the reduction in the letter contrast sensitivity of those with RP remains to be determined.

One important consequence of the overall loss of visual acuity from patients with RP is that their visual acuity impairment was greatest at the lowest contrast levels (Fig. 4). This relationship between log MAR and log contrast may account for our previous failure to obtain visual acuity measurements with Regan acuity charts in most such patients. This failure stems, not from a limited range of available letter sizes, but from the Regan chart approach, which measures log MAR at low levels of letter contrast. Our results indicate that, at contrasts below approximately 5%, it would be impossible to increase the size of the visual acuity targets sufficiently to enable subjects with RP to identify them correctly, although measurements could be made in control subjects under these conditions.

In addition to a disproportionately greater loss of visual acuity at low contrast levels, patients with RP have been observed previously to show a greater visual acuity loss at low luminances. An explanation for this finding can be derived from Figure 2. In this figure, the intersection of a set of LCSFs with the abscissa represents an acuity–luminance function obtained at a letter contrast of 1.0. For the control subjects, the increasing horizontal separation between the intersection points of the LCSFs at lower adapting levels shows that the visual acuities of these normal control subjects became progressively more impaired at lower luminances, as would be expected. A similar relationship between visual acuity and luminance was observed in Figure 2 for patients with RP. However, because of their overall loss of contrast sensitivity, this group had a proportionally greater separation between the intersection points at low luminances than did the control group. Consequently, the disproportionately greater loss of visual acuity in affected patients at low luminances may result from a reduction in the effective contrast of the visual acuity targets.

Additional understanding of the loss of visual acuity at low luminances can be derived from the letter decrement threshold functions. As indicated by the horizontal line segments in Figures 5 and 6, the minimum letter size that can be identified at a given adapting level (ie, visual acuity) is constrained by the...
maximum available luminance decrement. As adapting field luminance decreases, the maximum available luminance decrement also decreases. At the higher adapting field luminances, letter decrement thresholds decrease at approximately the same rate as the maximum available luminance decrement (Weber's law). Consequently, at the higher adapting levels, visual acuity tends to remain relatively constant as the background luminance decreases. However, at low adapting levels, where the decrement threshold functions are flatter, decreasing the adapting field luminance means that a smaller luminance decrement is available, and the decrement threshold for small letters becomes greater than the maximum available luminance decrement. Therefore, there is a decrease of visual acuity at low adapting levels because of physical constraints imposed by the use of negative-contrast letters; an explanation based on a loss of lateral inhibition is not necessary to account for this finding. A reduction in luminance does not alter the inhibitory properties of spatial frequency mechanisms. According to our results, then, visual acuity at low luminances could be improved if letters of positive rather than negative contrast were used.

In conclusion, the letter contrast sensitivities of this group of patients with RP were reduced below normal at all letter sizes and luminance levels. Specifically, their letter contrast sensitivity functions were displaced from normal horizontally, corresponding to an overall loss of contrast sensitivity. As a consequence, their foveal visual acuity losses were considerably greater at low contrasts and low luminances than under standard clinical testing conditions. The overall pattern of results was most consistent with the hypothesis that a general increase in foveal intercone spacing, rather than a reduced quantal absorption or a random loss of foveal cones, accounts for the abnormal letter contrast sensitivity functions of these patients with RP.

Key words: retinitis pigmentosa, contrast sensitivity, cones, acuity, adaptation, spatial vision

Acknowledgments

The authors thank Dr. Neal Peachey for helpful comments on the manuscript and Dr. Marlos Viana for statistical advice.

References