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The Relationship Between Visual Disability and Visual Scores in Patients With Retinitis Pigmentosa

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Purpose: To evaluate the relationship between visual disability and visual scores in patients with retinitis pigmentosa.

Methods: The relationship between visual disability and visual scores (visual acuity and visual field) was investigated in 93 patients with retinitis pigmentosa. The visual disability of each patient was evaluated using a questionnaire (a total of 35 questions, in 7 sections, regarding daily life). The reproducibility and validity of the data obtained by the questionnaire had been established by a similar investigation in glaucoma patients. Mean (±SD) age of patients was 52.6 ± 15.1 years, the mean visual acuity of the logarithm of the minimum angle of resolution ($\log_{10}$MAR) was 0.5 ± 0.4, and the mean deviation of visual field with the Humphrey Field Analyzer program 30-2 was −22.0 ± 10.9 dB.

Results: The visual acuity of $\log_{10}$MAR in the better eye ($r = 0.66$ to 0.81) and the mean sensitivity within the central 10° of the visual field ($r = -0.76$ to -0.62) had a definite relationship to the visual disability index of each section and their sum ($P < .0001$). This relationship was also confirmed in multiple regression analysis, which showed a high correlation coefficient ($R^2 = 0.57$ to 0.77, $P < .0001$).

Conclusions: The retinal sensitivity within the central 10° and the visual acuity of $\log_{10}$MAR in the better eye had a significant influence on a patient's daily life. We suggest that in patients with retinitis pigmentosa, visual disability in daily life can be precisely evaluated with the retinal sensitivity within the central 10° and the visual acuity in the logarithm of the minimum angle of resolution in the better eye. **Jpn J Ophthalmol 2000;44:82–87** © 2000 Japanese Ophthalmological Society

Key Words: Questionnaire, retinitis pigmentosa, visual acuity, visual disability, visual field.

Introduction

Retinitis pigmentosa (RP) is characterized by progressive photoreceptor and retinal pigment epithelium degeneration.[1] As the disease progresses, the degree of patients' subjective symptoms, such as visual field loss and central vision loss, increases and causes severe visual disability in their daily life. The relationship between visual disability and clinical assessment of visual functions, including visual field, visual acuity, or electroretinographic data in RP patients already has been reported[2–5]; however, only the visual field has been assessed by Szlyk et al.[3,4] Because they used the Goldmann perimeter for visual field testing, the central visual field that seems to have a great influence on daily visual function has not been assessed in their study. We have previously evaluated the relationship between visual disability and the central visual field with program 30-2 of the Humphrey Field Analyzer (HFA 30-2; Carl Zeiss, Dublin, CA, USA) in patients with glaucoma, which is regarded as one of the representative diseases, such as RP, which cause visual field impairment. We reported that the mean sensitivity within the central 10° (especially the lower hemifield within the central 5°) showed the strongest correlation with the visual disability in glaucoma patients.[6,7] In the present study, we evaluated the relationship between visual disability and the central visual field assessment by HFA 30-2 in patients with RP, whose pattern of visual field defect differs from glaucoma.

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Materials and Methods

Subjects

The degree of visual disability in daily life was determined in 93 patients (50 men and 43 women) with RP. The genetic type of RP was either autosomal recessive or isolated. The mean (± SD) age of patients was 52.6 ± 15.1 years. The mean best corrected visual acuity in the logarithm of the minimum angle of resolution ($\log_{10}$MAR) was 0.4 ± 0.4 in the eye with better vision (better eye) and 0.6 ± 0.4 in the eye with worse vision (worse eye) (Figure 1). The mean deviation of HFA 30-2 data was −21.0 ± 9.6 dB in the better eye and −23.5 ± 10.5 dB in the worse eye (Figure 2).

Patients who had visual impairment from other ocular diseases were excluded from this study.

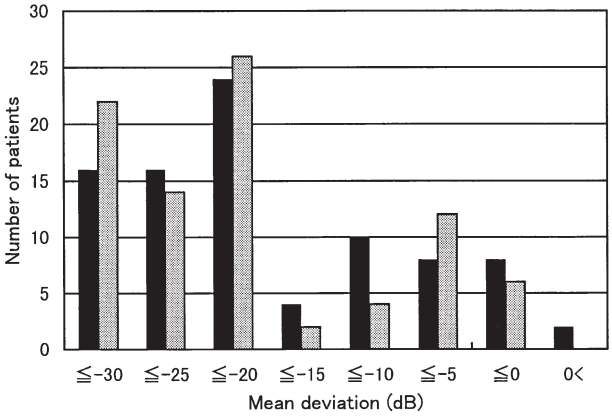

Figure 2. Mean deviation of Humphrey program 30-2 in subjects. Black, better eye. Gray, worse eye.

Methods

Questionnaire. The visual disability of each patient in daily life was evaluated using a questionnaire that was originally written in Japanese (Table 1). It contained a total of 35 questions in 7 sections—legibility of letters (letters), sentences (sentences), walking, going out by public transportation (going out), dining (dining), clothing and dressing (clothing), and others. The reproducibility and validity of the data obtained by the questionnaire had been established by a similar investigation in glaucoma patients.[6] In consideration of the difference between glaucoma and RP, each patient was also asked to write about visual disability in daily life, with respect to factors that were not covered in the questionnaire. Each question had three types of response choices, which were scored as follows: greatly disabled, 2 points; slightly disabled, 1 point; and not disabled, 0 point.

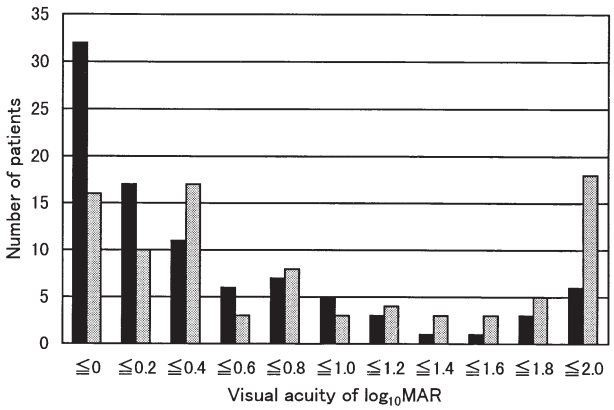

Figure 1. Best corrected visual acuity of $\log_{10}$MAR in subjects. Black, better eye. Gray, worse eye.

The visual disability index (DI) of each section (section DI: the mean score within each section) and their sum (total DI) were calculated for each patient.

Evaluating Visual Acuity

Binocular visual acuity was expected to have a relationship to visual disability. Because visual acuity differed in the right eye and the left eye in many of our subjects, we compared the relationship between visual disability and three representative visual acuity data—the visual acuity in the better eye, the visual acuity in the worse eye, and the mean visual acuity for both eyes (mean)—to determine which would best describe the visual disability in RP patients. This visual acuity was used in $\log_{10}$MAR in the statistical analysis. Pearson's correlation coefficient was used to assess the correlation between visual DIs (section DI and total DI) and individual visual acuity data.

Testing Visual Field

The central visual field was tested with HFA 30-2. The visual field index (VFI) within the central 30° (VFI 30) was calculated from the mean of representative sensitivity at each stimulus point within the central 30°. Comparing the sensitivity in the right and left eyes at each stimulus point: (1) higher sensitivity, (2) lower sensitivity, and (3) the mean sensitivity of both eyes were selected to represent sensitivity at each stimulus point when calculating VFI 30. Pearson's correlation coefficient was calculated between the visual DIs and VFI 30 in (1) to (3), above, to determine which method showed the strongest correlation with the visual DIs. Of the three methods

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Table 1. Questions Included in Questionnaire

Legibility of letters (Letters)	
Can you read the headlines in a newspaper?	(Yes/With difficulty/No)
Can you read small print in a newspaper?	(Yes/With difficulty/No)
Can you read letters in a dictionary?	(Yes/With difficulty/No)
Can you see numbers in a telephone directory?	(Yes/With difficulty/No)
Can you read a fare table for trains and subways?	(Yes/With difficulty/No)
Sentences	
Do you feel difficulty in reading and writing?	(No/Occasionally/Frequently)
When you write sentences in vertical lines, does it lean to either direction?	(No/Occasionally/Frequently)
Can you read movie subtitles?	(Yes/With difficulty/No)
When you read, can you find the next line easily?	(Yes/With difficulty/No)
Walking	
Do you feel difficulty in walking because of your visual problems?	(No/Occasionally/Frequently)
Can you take a walk by yourself?	(Yes/With difficulty/No)
Do you misjudge traffic signals?	(No/Occasionally/Frequently)
Do you bump into people or objects while walking?	(No/Occasionally/Frequently)
Do you stumble on the stairs?	(No/Occasionally/Frequently)
Do you fail to notice changes in ground level?	(No/Occasionally/Frequently)
Do you fail to recognize your friends until they talk to you?	(No/Occasionally/Frequently)
Do you fail to see people or cars approaching you from the side?	(No/Occasionally/Frequently)
Going out	
Do you feel difficulty in going out because of your visual problems?	(No/Occasionally/Frequently)
Do you need somebody to accompany you to go to a new place?	(No/Preferably/Yes)
Can you get a cab by yourself?	(Yes/With difficulty/No)
Do you have difficulty in traveling by train?	(No/Occasionally/Frequently)
Do you feel uneasy to go out at night because of your visual problems?	(No/Occasionally/Frequently)
Dining	
Do you feel difficulty in dining because of your visual problems?	(No/Occasionally/Frequently)
Can you dine by yourself?	(Yes/With difficulty/No)
Do you drop food while dining because of your visual problems?	(No/Occasionally/Frequently)
Do you spill tea while pouring into a cup?	(No/Occasionally/Frequently)
Do you feel difficulty in using chopsticks?	(No/Occasionally/Frequently)
Clothing and dressing (Clothing)	
Can you change clothes by yourself?	(Yes/With difficulty/No)
Do you sometimes put underwear on inside out?	(No/Occasionally/Frequently)
Do you ever button up clothing in the wrong order?	(No/Occasionally/Frequently)
Do you have difficulty dressing because of your visual problems?	(No/Occasionally/Frequently)
Can you see your face clearly in the mirror?	(Yes/With difficulty/No)
Others	
Can you recognize people's faces on TV?	(Yes/With difficulty/No)
Do you have difficulty finding objects dropped on the floor?	(No/Occasionally/Frequently)
Do you have difficulty dialing the telephone unless you look very closely?	(No/Occasionally/Frequently)

of calculating VFI 30, the method that showed the strongest correlation with the visual DIs was adopted in the following statistical analysis. To decide which section of the visual field within the central 30° has the strongest correlation with visual DIs, the VFIs from 0° (point of fixation) to 10° (VFI 10), from 11° to 20° (VFI 11–20), and from 21° to 30° (VFI 21–30) were calculated. The visual field area having a representative sensitivity of 20 dB or better, corresponding to the I-4-e target of the Goldmann perimeter[8] (Area-20 dB), was also calculated in each patient. Area-20 dB was defined as the sum of the degrees of the visual field from the point of fixation at the eight principal meridians (up, up and nasally, nasally, down and nasally, down, down and temporally, temporally and up, and temporally) within the central 30°.

Pearson's correlation coefficient was used to assess the relationships between the visual DIs and the VFI 10, VFI 11–20, VFI 21–30, and Area-20 dB.

Table 2. Pearson's Correlation Coefficients Between Visual Acuity Shown in $\log_{10}$MAR and Visual Disability Indices

	Section Disability Index (DI)							
Visual Acuity	Letters	Sentences	Walking	Going Out	Dining	Clothing	Others	Total DI
Better eye	0.74*	0.71*	0.66*	0.75*	0.74*	0.74*	0.80*	0.81*
Worse eye	0.67*	0.62*	0.56*	0.65*	0.63*	0.61*	0.72*	0.70*
Mean	0.70*	0.62*	0.54*	0.66*	0.66*	0.66*	0.73*	0.73*

*$P < .0001$.

Multiple Regression Analysis

Multiple regression analysis was also used to assess the correlation between each dependent variable (the visual DIs: section DI and total DI) and independent variables (patient history [sex, age, and occupation], visual acuity [in the better eye and in the worse eye], and visual field [VFI 30, VFI 10, VFI 11–20, VFI 21–30, and Area-20 dB]). Independent variables were selected for each dependent variable by the stepwise variable selection method ($P < .05$). Standard partial regression coefficients for each independent variable and R^2 value were calculated.

Results

As shown in Table 2, visual acuity in the better eye, the worse eye, and the mean of both eyes correlated strongly with the DIs. Especially, the visual acuity in the better eye showed the strongest correlation with all the DIs ($r = 0.66$ to 0.81, $P < .0001$).

As shown in Table 3, regardless of which method was used for calculating the representative sensitivity within the central 30° in the HFA 30-2 assessment, there was a significant correlation with the DIs. However, when we adopted the higher sensitivity as the representative sensitivity at each stimulus point, the strongest correlation was shown ($r = -0.64$ to -0.43, $P < .0005$). Therefore, we adopted the higher sensitivity as the representative sensitivity of each stimulus point when calculating the VFI 10, VFI 11–20, VFI 21–30, and Area-20 dB. The VFI 10, VFI 11–20, VFI 21–30, and Area-20 dB also showed significant correlation with the DIs, and of these, the VFI 10 showed the strongest correlation with all DIs ($r = -0.76$ to -0.62, $P < .0001$). There was no significant correlation between patient age and any of the DIs.

Table 3. Pearson's Correlation Coefficient Between Visual Field Measured by HFA 30-2 and Visual Disability Indices

	Section Disability Index (DI)							
HF30-2	Letters	Sentences	Walking	Going Out	Dining	Clothing	Others	Total DI
VFI 30								
Higher*	−0.43‖	−0.49‖	−0.64§	−0.63§	−0.50‖	−0.48‖	−0.55§	−0.59§
Lower†	−0.41#	−0.46‖	−0.58§	−0.58§	−0.46‖	−0.45‖	−0.50‖	−0.55§
Mean‡	−0.41#	−0.47‖	−0.60§	−0.60§	−0.47‖	−0.45‖	−0.51‖	−0.56§
VFI 10	−0.62§	−0.66§	−0.68§	−0.76§	−0.68§	−0.63§	−0.76§	−0.76§
VFI 11–20	−0.43‖	−0.50‖	−0.64§	−0.59§	−0.48‖	−0.45‖	−0.57§	−0.58§
VFI 21–30	−0.32††	−0.42¶	−0.56§	−0.47‖	−0.40#	−0.33**	−0.43‖	−0.46‖
Area-20 dB	−0.40#	−0.47‖	−0.44‖	−0.50‖	−0.43‖	−0.41#	−0.50‖	−0.50‖

*Higher sensitivity.
†Lower sensitivity.
‡Mean sensitivity of both eyes used to calculate representative sensitivity at each stimulus point.
§$P < .0001$.
‖$P < .0005$.
¶$P < .001$.
#$P < .005$.
**$P < .01$.
††$P < .05$.

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Table 4 contains the standard partial regression coefficients and R^2 values of the multiple regression analysis. For all DIs the independent variables selected by the stepwise variable selection method were visual acuity in the better eye and the VFI 10 obtained with HFA 30-2. No other independent variables were selected for any of the DIs. R^2 values between the predicted scores and the actual scores of section DI and total DI were 0.57 to 0.77 and 0.75, respectively ($P < .0001$).

Patients were asked to write about factors that affected visual disability in daily life that were not included in the questionnaire, four items (walking down steps in dark places, going out in the twilight, walking on a bright sunny day, and reading books at night) were reported. They were all included in the questionnaire.

Discussion

The relationship between visual disability in daily life and the clinical assessment of visual acuity and visual field with the HFA 30-2 were evaluated in RP patients. The visual acuity of $\log_{10}$MAR in the better eye and the mean sensitivity within the central 10° of the visual field had a definite relationship to the visual DIs.

In Japan, visual disability in patients with RP has already been reported by Hayakawa et al[2] in what they called a "Quality of Life Questionnaire," based on a preliminary survey of 151 patients. The questionnaire included the patient's state of mind and social activity evaluations, in addition to eight items concerning daily life functions, such as walking, going out, and the ability to identify traffic signals. To evaluate the overall satisfaction level of each patient, they categorized the degree of visual disability into two levels. However, they did not report the relationship between the clinical assessment of visual functions (such as visual acuity and visual field) and the visual disability.

In the United States, Szlyk et al[3] assessed visual disability in daily activities in RP patients using a questionnaire modified from "The Activities of Daily Vision Scale,"[9] which was originally developed as a means of evaluating visual function in patients with cataracts. They also added questions intended to target potentially problematic activities specifically for RP patients, for example, going out at night, shopping, stepping on and off escalators, and finding a seat in dark movie theaters. They assessed the correlations between the visual disability and the clinical assessment of visual function, including visual acuity, electroretinographic data, and visual field area measured by the Goldmann perimeter. The perimeter of an eye was selected randomly, and visual field data measured by static perimeter were not assessed.

The questionnaire used in the present study is formulated on the basis of a preliminary survey for patients with glaucoma; its validity and reproducibility have already been reported.[6] Our questionnaire does not include survey items involving cooking, cleaning, reading price tags, etc., as appeared among the survey items of Hayakawa et al.[2] Neither does it include the survey items of Szlyk et al,[3,4] such as driving a car, using an escalator, and playing cards. On the other hand, our survey item concerning the patient's visual disability in changing clothes was not

Table 4. Standard Partial Regression Coefficient and R^2 Value in Multiple Regression Analysis

	Standard Partial Regression Coefficient (Standard Error)		
Dependent Variable	Visual Acuity in Better Eye	VFI 10	R^2 value
Section disability index			
Letters	0.50 (0.14)	−0.52 (0.01)	0.60*
Sentences	0.37 (0.007)	−0.40 (0.14)	0.62*
Walking	0.37 (0.006)	−0.46 (0.11)	0.57*
Going out	0.41 (0.11)	−0.47 (0.006)	0.71*
Dining	0.44 (0.09)	−0.33 (0.004)	0.63*
Clothing	0.51 (0.08)	−0.26 (0.004)	0.60*
Others	0.46 (0.005)	−0.49 (0.009)	0.77*
Total disability index	0.47 (0.59)	−0.41 (0.03)	0.75*

VFI: Visual field index.
*$P < .0001$.

included in the studies of either Hayakawa et al or Szlyk et al. Before we compiled our questionnaire, many patients with glaucoma complained of difficulty with changing clothes. Therefore, changing clothes was considered one of the important items of our questionnaire. This item is also included in "The Visual Functioning Index,"[10] which is the questionnaire for evaluating visual disability in daily life in patients with cataracts, while the same item was also incorporated into "The 36-Item Short-Form,"[11] which is one of the representative questionnaires for evaluating quality of life.[12] It is evident that the answers to the questionnaires will not only reveal information about the level of disability, which may be subjective, but will also reflect the lifestyle of each patient, ie, habits, etc., as well as cultural factors. Our subjects felt that no factors had been omitted in our questionnaire that related to their visual disability, as was revealed by the statements and declarations that we elicited from them.

In a fashion similar to our present study, Szlyk et al[3] have already reported that visual acuity and the visual field area of the II-4-e target of the Goldmann perimeter were significantly related to visual disability. However, Szlyk et al adopted visual acuity for either the right eye or the left eye randomly, and failed to compare the worse-eye data with the better-eye data in the same patient. In the evaluation of the relationship between visual disability and visual acuity in patients with cataracts, it has been reported that it was the visual acuity in the better eye that had the greatest bearing on visual disability. In our study, we obtained similar results from patients with RP. The correlation coefficient between each visual DI and visual acuity in the better eye was even higher than that of Szlyk et al. From these findings, we considered that the evaluation of visual acuity in the better eye is appropriate for evaluating visual disability in patients with RP.

Other findings in our present study revealed that the relationship between visual disability in daily life had a more significant correlation with the mean retinal sensitivity within the central 10° than with a visual field area having the retinal sensitivity of 20 dB or better. The correlation coefficient between the visual field area of 20 dB or more and visual disability was 0.40 to 0.50, which was more than that of 0.10 to 0.45 of the visual field area measured by the II-4-e target of the Goldmann perimeter in the report of Szlyk et al. In our study the mean retinal sensitivity within the central 10° shows further improvements of 0.62 to 0.76, which agrees with the results in our previous study in patients with glaucoma.[6] This result indicated that retinal sensitivity within the central 10° had a significant influence on a patient's daily life.

Moreover, the multiple regression analysis of our study showed that, among patient's sex, age, occupation, visual acuity, the mean retinal sensitivity within each section, and the visual field area of 20 dB or more, only the mean sensitivity within the central 10° and the visual acuity in the better eye showed a significant correlation with visual disability. Therefore, we suggest that, in patients with RP, visual disability in daily life can be precisely evaluated on the basis of the mean sensitivity within the central 10° and the visual acuity in the better eye.

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