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Assessment of Functional Vision Score and Vision-Specific Quality of Life in Individuals With Retinitis Pigmentosa

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Purpose: To determine the relationship between the American Medical Association's (AMA) functional vision score (FVS) and vision-specific quality of life in retinitis pigmentosa (RP) patients using the National Eye Institute's Visual Functioning Questionnaire (NEI-VFQ 25).

Methods: One hundred eight patients with RP participated in the study. We measured best-corrected visual acuity, conducted Goldmann perimetry, and collected the self-reported NEI-VFQ 25. The FVS was calculated using the functional field score (FFS) and the functional acuity score (FAS). The correlations of the VFQ composite scores to the FVS, FFS, and FAS were determined using correlation and regression analyses.

Results: FVS was highly correlated to the BCVA (r=0.69, p<0.001), the FFS (r=0.86, p<0.001) and the FAS (r=0.73, p<0.001). Significant correlations of the VFQ composite score to the BCVA (r=0.60, p<0.001), FFS (r=0.44, p<0.001), FAS (r=0.60, p<0.001), FVS (r=0.58, p<0.001) were also found. However, the correlation strengths of BCVA, FVS, FAS, and FFS to NEI-FVQ were not different.

Conclusions: In RP patients, the vision-specific quality of life was correlated with the AMA guidelines' FVS, FFS, and FAS. Their correlation degrees to NEI-FVQ were not different. This result suggests that vision-specific quality of life can be explained by both visual acuity and visual field in RP patients.

Key Words: AMA guidelines, Functional acuity score, Functional field score, Functional vision score (FVS), NEI-VFQ 25, Retinitis pigmentosa

Retinitis pigmentosa (RP), which leads to retinal degeneration, is characterized by nyctalopia, intraretinal bony specule pigmentation, vessel attenuation, rod-cone dysfunction as determined by electroretinogram and progressive visual field loss leading to blindness.[1] Impaired vision can be evaluated by measuring visual acuity (VA) or by visual field test.[2] However, these measurements alone are of limited value in evaluating vision-specific quality of life; hence, several studies[3-6] have been conducted to document visual function and measure performance in RP patients. Recently, the National Eye Institute's Visual Functioning Questionnaire (NEI-VFQ 25) composite scores were suggested for evaluating vision-specific quality of life, and the reliability of this method was proved in several studies for chronic diseases (glaucoma, ARMD).[7,8] The previous studies, however, did not evaluate the relationship between vision-specific quality of life and the American Medical Association (AMA) guidelines' functional vision score (FVS) in RP patients. The AMA guidelines' FVS was reported to be a better predictor of vision-targeted quality of life than traditional measurements of visual acuity or visual field extentin disease.[9] However, it still remains unclear whether the vision-specific quality of life correlates with objective visual measurements in retinal diseases such as RP.

In this study, we measured the best-corrected visual acuity (BCVA) and visual field extent while collecting data from the self-reported NEI-VFQ 25 from RP patients to determine the relationship of the FVS and vision-specific quality of life. We also analyzed the effect of each visual measurement to the quality of life in order to evaluate the importance of each visual scale in RP patients at different clinical stages. To our knowledge, our study offers the first report of the correlation of FVS to vision-specific quality of life in RP patients using the NEI-VFQ 25.

Materials and Methods

We enrolled 108 volunteers (65 males, 43 females) with RP, ranging in age from 16 to 85 years, who were members of the Korean Retintis Pigmentosa Society. A Korean national RP survey was conducted at Seoul National University Hospital's retinal clinic from July to December 2006. RP was diagnosed in the recruited patients on the basis of a fundus examination, Goldmann perimetry, and a complete electroretinographic

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Table 1. Classification of functional vision score[11]

Class	Description	Estimated ability to perform activities of daily living	FVS (points)
1	Range of normal vision	Retained reserve capacity	>90
2	Near-normal vision	Lost reserve capacity	71-90
3	Moderately low vision	Need for visual enhancement of aids	51-70
4	Severely low vision	Slower than normal, even with enhancement aids	31-50
5	Profoundly low vision	Marginal visual performance, even with aids	11-30
6	Total blindness	Cannot perform, needs substitution aids	<10

FVS=functional vision score according to AMA guides.
Source: Guides to the evaluation of permanent impairment, fifth edition, copyright 2001, American Medical Association.

Table 2. Demographic and clinical characteristics of enrolled patients (n=108)

Variables	Mean±SD (Range)	Variable	n, (percent)
Age (years)	37.6±13.0 (13.0-85.0)	Gender (male %)	65, (60.1 %)
Disease duration (years)	19.1±12.9 (1-58)	FVS Class distribution	4.9±0.8 (1-6)
Onset age (years)	19.0±14.9 (1-80)	1	0, (0 %)
BCVA OD	0.81±0.68 (0.0-2.3)	2	0, (0 %)
OS	0.78±0.68 (0.0-2.3)	3	5, (4.6 %)
BCVA better eye	0.63±0.58 (0.0-2.3)	4	23, (21.3 %)
FVS	21.3±16.1 (0.0-68.5)	5	50, (46.3 %)
FFS	29.7±17.1 (0.8-69.2)	6	30, (18.5 %)
FAS	65.3±27.9 (0.0-100.0)		
NEI-VFQ			
General vision	37.2±17.6 (0.0-80)		
Ocular pain	71.3±24.9 (0.0-100)		
Near vision	55.6±29.3 (8.3-100)		
Distant vision	49.2±22.7 (0.0-91.7)		
Vision-specific social function	61.9±26.3 (12.5-100)		
Vision-specific mental health	42.3±26.2 (0.0-100)		
Vision-specific role difficulty	39.7±30.5 (0.0-100)		
Vision-specific dependency	58.6±27.0 (0.0-100)		
Driving	57.5±28.9 (0.0-100)		
Color vision	72.5±30.7 (0.0-100)		
Peripheral vision	46.3±25.8 (0.0-100)		
Composite score	50.8±19.8 (14.6-95.9)		

FVS=functional vision score; FFS=functional field score; FAS=functional acuity score; NEI-FVQ=National Eye Institute's Visual Functioning Questionnaire.

evaluation according to the parameters of the International Society for Clinical Electrophysiology of Vision (ISCEV). Patients with hearing impairment (Usher syndrome) or other systemic diseases were excluded. However, patients with reading difficulties due to low vision were not excluded. The researchers assisted these patients in completing the questionnaires. Patients were given the option of dropping out of the study. Our Institutional Review Board (IRB) approved the study protocol, informed consent was obtained from all of the subjects, and all procedures used were consistent with the tenets of the Helsinki Declaration. All of the patients underwent a thorough ophthalmic examination including best-corrected visual acuity (BCVA), binocular indirect ophthalmoscopy, fundus examination, and Goldmann perimetry. If a definite diagnosis of RP could not be made, a standard electroretinogram was performed for confirmation.

Visual acuity measurement and visual field examination

The BCVA was measured using Snellen Visual Acuity Charts and was converted into a logarithm of the minimum angle of resolution (log MAR) VA scale. Monocular visual fields were measured by Goldmann perimetry using the Ⅲ-4-e target at a standard luminance. Along each meridian, the target was presented from a position of non-seeing to seeing, moving clockwise. All of the BCVA and perimetry measurements were performed by skilled technicians.

Functional assessment according to the guidelines

The AMA has published guidelines[10,11] for the evaluation of permanent impairment. The FVS was calculated from the functional acuity score (FAS) and the functional field score

Table 3. Correlation of the American Medical Association functional vision scores to retinitis pigmentosa patients

	FVS	BCVA	FAS	FFS
BCVA	r=0.69, p<0.001			
FAS	r=0.73, p<0.001	r=0.94, p<0.001		
FFS	r=0.86, p<0.001	r=0.38, p<0.001	r=0.39, p<0.001	
NEI-VFQ	r=0.58, p<0.001	r=0.60, p<0.001	r=0.60, p<0.001	r=0.44, p<0.001

BCVA=best-corrected visual acuity; FVS=functional vision score; FFS=functional field score; FAS=functional acuity score; NEI-VFQ =composite score.
Spearman correlation analysis, two-sided p-values of <0.05 were considered statistically significant.

Table 4. Comparison of functional acuity score and functional field score between the better visual acuity group and worse visual acuity group

Variables	Better VA group (n=46)	Worse VA group (n=62)	p-value*
Age (years)	34.7±9.7	39.8±14.7	0.03
Gender (male %)	35, 76 %	30, 48.2%	0.84
Disease duration (years)	16.6±12.5	20.8±13.0	0.13
Age of onset (years)	17.8±10.2	19.8±17.4	0.49
BCVA OD	0.41±0.56	1.09±0.60	<0.001
OS	0.31±0.37	1.13±0.64	<0.001
BCVA, better eye	0.18±0.13	0.97±0.55	<0.001

FVS=functional vision score; FFS=functional field score; FAS=functional acuity score; VA=visual acuity.
* Independent t-test, two-sided p-values of <0.05 were considered statistically significant.

(FFS), as defined in the aforementioned guidelines.[10,11] VA measurements were converted to a visual acuity score (VAS). The weighted average of three VASs for each field was used to calculate the FAS according to: FAS=(VAS_{OD}+VAS_{OS}+3×VAS_{OU})/5.

To evaluate the FFS, the visual field score (VFS) for the right monocularfield (VFS_{OD}), the left monocular field (VFS_{OS}), and the binocular field (VFS_{OU}) were first scored separately: FFS=(VFS_{OD}+VFS_{OS}+3×VFS_{OU})/5.

The FAS and FFS were then multiplied to yield the FVS: FVS=FAS×FFS/100.

The AMA FVS classification (Table 1) was used to classify patients.[11]

Self-Reported Questionnaire (NEI-VFQ 25)

The NEI-VFQ 25-item version with appendix[7,8] (a total of 39 items) was administered by skilled interviewers and scored in the standard manner. There were twelve sub-scale scores and one composite score. The NEI-VFQ 25 composite score was the average of all available sub-scales, except general health, and was suggested as the vision-related quality of life indicator by the NEI.

Statistical analysis

The correlations of the NEI-VFQ 25 composite score to the FVS, FFS, and FAS were analyzed by the Spearman correlation test. If the correlations were significant, Fisher's Z-transformation analysis was used to determine the better predictor of vision-specific quality of life among the FVS, FFS, and FAS. A regression analysis was performed to determine regression equations. As the median log MAR was 0.6, we divided the patients into two groups according to that value: the better VA group (logMAR<0.6), and the worse VA group (logMAR≥0.6). In each group, the relationship of theVFQ composite score to the FVS, FFS, and FAS was evaluated and regression analysis was performed. Statistical analyses were performed using SPSS v.12.0 software (SPSS Inc., Chicago, IL), and two-sided p-values of <0.05 were considered statistically significant.

Results

As stated above, there were 108 RP patients enrolled (65 males, 43 females) ranging in age from 16 to 85 years. The mean age of the subjects was 37.6±13.0 years. Their VA ranged from 0.0 to 2.3 log MAR. The demographics and descriptive statistics for the clinical measures of visionare listed in Table 2. Most participants were classified into more advanced categories (FVS 4, 5, 6).

FVS was highly correlated to the BCVA (r=0.69, p<0.001), FFS (r=0.86, p<0.001) and the FAS (r=0.73, p<0.001) (Table 3). Significant correlations of the VFQ composite score to BCVA (r=0.60, p<0.001), FFS (r=0.44, p<0.001), FAS (r=0.60, p<0.001), FVS (r=0.58, p<0.001) were also found. However, we could not find any differences among the correlations of BCVA, FVS, FFS, and FAS to the VFQ composite score. Study patients were older in the poor VA group than in the better VA group (34.7±9.7 vs. 39.8±14.7, p=0.03, Table 4). Multiple regression analysis was performed as follows with the interactive forms of FFS and FAS.

(All patients, n=112), NEI-FVQ composite=0.30×FAS+

0.31×FFS+21.72, (r^2=0.40).
(Better VA group, n=50), NEI-FVQ composite=1.50×FAS +2.91×FFS-2.78×FVS-86.11 (r^2=0.37).
(Worse VA group, n=62), NEI-FVQ composite=0.24×FAS +0.23×FFS-2.78+25.8 (r^2=0.22).

Discussion

Our results indicate that in RP patients, BCVA and the AMA guidelines' FVS, FFS, and FAS are equally correlated to those of the self-reported VFQ. Several other studies[6,12] evaluating the performance of RP patients demonstrated that reading performance correlates with contrast sensitivity, VA, and visual field, while driving performance is the primary correlate of visual field loss. In fact, one of the conclusions of the work is that FVS is no better than BCVA (correlation: 0.60) for categorizing RP patients in terms of self-perceived QOL. This can be expected since the visual functioning of RP patients can be estimated from BCVA on average, although contrast sensitivity may add information.

To assess performance function, several studies[7-9] utilized questionnaires or AMA guidelines for FVS. According to the results, the VFQ is a reliable, valid method that should be useful for group-level comparisons of vision-specific quality of life in clinical research. The FVS has also been found to be a potent predictor of self-reported vision-specific quality of life. However, these studies[9,13,14] did not focus on RP patients. Our study confirmed that the BCVA, FVS, FAS and FFS are highly correlated to the VFQ in RP patients. In accordance with our results, Szlyk et al. found that self-reporting is strongly correlated with actual task performancein RP patients.[5] The group evaluated the correlation of reading composite scores with contrast sensitivity, whereas, in our study, we used the AMA's FVS and VFQ composite score. However, compared with BCVA and FVS, it suggests that the FVS may not add much to the value of basic measures of visual function in some diseases. We could comment on previous research on the FVS in other diseases or in the general population, such as the study by Rubin et al.[15] who stated that monocular acuity and binocular acuity are significantly better predictors of reading speed than the AMA weighted score or a recently proposed Functional Vision Score (FVS).

In this study, the VFQ was affected by both the FFS and the FAS, although the regression equation had an interactive form (FVS). We speculate that these findings might reflect the fact that RP is a disease manifesting with progressive visual field loss. A hallmark feature of RP is an insidious, progressive loss of peripheral visual field. The peripheral island of visual field is lost before the central visual field contracts. Therefore, the remaining, functional field is important in advanced RP patients, as FFS represents visual field in the worse VA group. However, we cannot estimate the rate of visual field progression from our results, though Berson et al. suggested that the visual field is lost at a rate of about 4.6% per year.[16] Massof et al. proposed that the visual field diminishes approximately 50% over 4.5 years.[17] In any case, the rate of progression of visual field loss is usually slow and relentless in RP patients.

The limitations of this study include its cross-sectional design, which does not allow for the assessment of the RP course. Because the enrolled patients presented with various stages of RP, selection bias could be an issue. Despite these limitations, our study is the first to determine the correlation of FVS to vision-specific quality of life in a relatively large group of RP patients.

In conclusion, the vision-specific quality of life correlated with the AMA's guidelines with FVS, FFS and FAS in RP patients. The correlations to the NEI-FVQ were not different. These results suggest that visual quality cannot be explained only by visual acuity or visual field in RP patients.

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