Original Article

Vision-Related Quality of Life in Patients with Inherited Retinal Dystrophies

Saeideh Shojaei[1], Hamideh Sabbaghi[2], Yadollah Mehrabi[1], Narsis Daftarian[3], Koorosh Etemad[1], Hamid Ahmadieh[4]

[1]Department of Epidemiology, School of Public Health and Safety, Shahid Beheshti University of Medical Sciences, Tehran, Iran, [2]Ophthalmic Epidemiology Research Center, Research Institute for Ophthalmology and Vision Science, Shahid Beheshti University of Medical Sciences, Tehran, Iran, [3]Ocular Tissue Engineering Research Center, Research Institute for Ophthalmology and Vision Science, Shahid Beheshti University of Medical Sciences, Tehran, Iran, [4]Ophthalmic Research Center, Research Institute for Ophthalmology and Vision Science, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Abstract

Purpose: To evaluate the effect of inherited retinal dystrophies (IRDs) on vision-related quality of life (VRQoL) among IRDs' patients in Iran.

Methods: This cross-sectional study was conducted on 192 patients with different types of IRDs who were randomly selected from registered patients in the Iranian National Registry for Inherited Retinal Dystrophy (IRDReg®). All ophthalmic findings were collected based on the recorded data in IRDReg®. Moreover, the eligible participants were interviewed to fill out the National Eye Institute Visual Functioning Questionnaire-25 (NEI VFQ-25) to assess their VRQoL. Ordinal logistic regression was used to evaluate the possible association of the different clinical and nonclinical factors such as demographic information, socioeconomic status, and visual function with VRQoL.

Results: The overall mean of a composite score of VRQoL was 45. All subscales obtained from the NEI VFQ-25 questionnaire except general health, mental health, and ocular pain had a significant negative correlation with logMAR best corrected visual acuity (BCVA) and near visual acuity variables. There was a statistically significant relationship between VRQoL and factors like age (odds ratio [OR] = 0.91, 95% confidence interval [CI]: 0.87–0.94), employment status (OR = 1.37, 95% CI: 1.05–4.74), logMAR BCVA (OR = 0.31, 95% CI: 0.19–0.49) and normal color vision (OR = 1.92, 95% CI: 1.74–5.01).

Conclusion: The VRQoL of patients with IRDs in this study was low. BCVA could be an indicator to show VRQoL.

Keywords: Inherited retinal dystrophy, National eye institute visual functioning questionnaire-25, Vision-related quality of life

Address for correspondence: Koorosh Etemad, Department of Epidemiology, School of Public Health and Safety, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
E-mail: etemadk@gmail.com
Submitted: 30-May-2021; **Revised:** 16-Oct-2021; **Accepted:** 20-Oct-2021; **Published:** 16-Apr-2022

Introduction

Inherited retinal dystrophies (IRDs) are a group of rare ocular diseases caused by the gradual degeneration of photoreceptors, retinal pigment epithelium, and other retinal neurons leading to progressive vision loss and eventually blindness. Up to now, more than 300 genes have already been identified for inherited retinal diseases.[1,2]

The prevalence of IRDs is about 1 in 2000–3000 individuals, and it is estimated that over one million people may be affected worldwide. Epidemiologic studies show that the variation of IRDs diseases' prevalence directly depends on various demographic characteristics, including consanguinity, ethnicity, age, and the living area.[3,4] In this regard, retinitis pigmentosa (RP) and Leber's Congenital Amaurosis (LCA)

Access this article online

Quick Response Code:

Website: www.jcurrophthalmol.org

DOI: 10.4103/joco.joco_172_21

How to cite this article: Shojaei S, Sabbaghi H, Mehrabi Y, Daftarian N, Etemad K, Ahmadieh H. Vision-related quality of life in patients with inherited retinal dystrophies. J Curr Ophthalmol 2022;34:80-6.

are the most frequent types of IRDs, with the estimation of 1 in 4000 and 1 in 50,000–100,000 individuals, respectively.[5,6]

IRDs are reported as one of the leading causes of irreversible blindness which RP as the most prevalent type of IRDs is the leading cause of hereditary blindness in people younger than 60 years old.[5,7] IRDs are chronic and incurable, and they may affect both quality of life (QoL) and health economics by affecting physical well-being, psychological state, and productivity.[8,9]

In this regard, vision-related QoL (VRQoL) is defined as the patients' subjective perception, daily activities related to the visual ability, as well as social and emotional aspects of life related to the visual status.[10] Visual problems due to the IRDs resulted in decreased vision and the restriction of the visual field (VF), which directly affect the individual daily activities.[11]

The National Eye Institute Visual Function Questionnaire-25 (NEI VFQ-25) was developed recently from the NEI VFQ-51 to test the psychometric effects of visual impairment.[12] The best instrument to assess VRQoL is NEI VFQ-25. It is even more suitable than NEI VFQ-55 based on the length of the test and its feasibility while keeping the same comprehensiveness.[13] This questionnaire with 25 questions was reported to provide reliable and valid scores from patients with various eye diseases.[14-16]

There is no comprehensive information regarding VRQoL among Iranian patients who suffer from different types of IRDs. Only one study was performed on patients with RP that showed that mean NEI VFQ-25 scores were low in 35 RP patients, and decreased visual acuity (VA) has a great impact on the QoL.[17] However, in other countries, more extensive studies were performed on these patients. For instance, a qualitative study of IRDs' patients in the United States, Germany, and France showed that night blindness, reduced peripheral vision, and problems with color vision were the most frequent problems in these patients, and severity of limitations vary based on lighting conditions and familiarity of environment. These problems affect health-related QoL.[18] Prokofyeva *et al*. conducted a study on IRDs patients and showed that IRDs affect VRQoL.[19] Therefore, we proposed to do a national study of the VRQoL among these patients who lived in Iran. We also desired to investigate the relation of VRQoL in these patients with demographic, socioeconomic factors, and visual functions.

Methods

This is a retrospective study. The study population was the patients who suffered from different types of IRDs and were registered into the Iranian National Registry for Inherited Retinal Diseases (IRDReg®)[20] from January 2016 to December 2020. Information about ophthalmic tests, demographics variables, age of onset of IRD, and family history were extracted from the IRDReg®, and information about the QoL, and socioeconomic status (SES) were collected at the time of the study. All patients registered in the IRDReg routinely visit clinics for examinations every 6 months, and their information about best corrected visual acuity (BCVA), near VA, color vision, and ophthalmic examinations are updated.

A sample size of 192 was calculated, but it was planned to be increased to 225 individuals with consideration to the attrition rate of 15%. These patients were randomly selected from IRDReg®.

The ethics committee approved all Shahid Beheshti University of Medical Sciences study procedures via the approval number of IR.SBMU.PHNS.REC.1399.055. Informed consent was obtained from all study participants, and we assured the confidentiality of their information.

Only patients with a definite clinical diagnosis of IRDs by a retina subspecialist were included. We only interviewed patients with IRD aged over 15 years who were able to present a valid response. Patients with other ocular pathologies and/or systemic disabilities were not investigated in our analysis. In the patients with Usher syndrome who had hearing difficulty, data collection was conducted with the help of their family members or caregivers.

First, a list of all registered patients who were diagnosed with IRDs was received from IRDReg®. The list was sorted by file number, and patients who were registered more than once and individuals under 15 years old were deleted. Then using the RND function in Excel software, 225 people were selected by simple random sampling until reaching the sample size (192 people). Patients were contacted by telephone, and NEI VFQ-25 questionnaire was completed. The data of ophthalmic tests, demographics variables, age at symptom onset, duration of disease and family history were extracted from patients' documents in the IRDReg®, and information about VRQoL and SES were collected at the time of study by telephone. Finally, the data were merged and prepared for analysis.

Data were collected using a structured questionnaire translated into Persian, which consisted of questions for demographic factors and SES; VRQoL of participants with IRDs. NEI VFQ-25 including 12 subscales: General health, general vision, ocular pain, near activities, distance activities, social functioning, mental health, role difficulties, dependency, driving, color vision, and peripheral vision. The values were recorded on a 0–100 scale and averaged to generate scores for each subscale. The composite score was calculated as the average of all questions except general health (2 questions). Demographic factors and SES included age, sex, marital status, education, occupation, income, residency condition (rural or urban), floor area per capita, ethnicity, consanguinity, and age of symptom onset. NEI VFQ-25 questionnaire was assessed for reliability and validity by Asgari *et al*., and it was shown to accurately measure VRQoL in Iranian people.[21]

All IRDs patients underwent a comprehensive clinical examination, including assessing the corrected distance

visual acuity (CDVA) using Snellen E-chart. For the measurement of VA in this group of patients, we initially presented the vision chart to the patient at a distance of 3 m. Then the chart was moved to the patients upon the patient reporting to see the line. Distance reduction was carried out for one meter in each step up to the distance of 50 cm near the patient, and the VA was recorded based on the standard calculation. Afterward, we checked the qualitative assessment of counting finger, hand motion, and light perception only in cases that were not able to see the chart. Furthermore, color vision testing was performed using the Ishihara pseudoisochromatic 38-plates, and the evaluation of anterior and posterior segments was done by the biomicroscopy and indirect ophthalmoscope, respectively. All clinical information was completed based on the registered data in the IRDReg® database registry.

The composite score of VRQoL was set at 0–100 points in which 0 represented the worst and 100 represented the highest QoL. This composite score was converted to an ordinal variable (4 levels) based on quartile, which was assigned a score of 0–24.9, 25–49.9, 50–74.9, and 75–100.

VA classification was considered as presented in the International Classification of Diseases 11, so that IRDs patients with CDVA worse than 6/12–6/18 were classified as mild low vision, and those with CDVA in the range from 6/18–6/60 and 6/60–3/60 were considered as moderate and severe low vision, respectively. Furthermore, patients with CDVA worse than 3/60 were defined as blind.[22,23] Moreover, the color vision status was recorded as normal in patients who were able to respond to eight plates correctly. Otherwise, they were considered color vision deficient.[24] The test results in the patients with no perception of demo plate were also recorded as not applicable (NA).

Demographic and socioeconomic variables used in this study were collected face-to-face or by telephone interview along with NEI VFQ-25 questionnaires. Demographic and SES variables, including age, incidence age, duration of illness, sex (male, female), marital status (single, married), level of education (under diploma, diploma, academic), occupation status (unemployed, employed), level of parental consanguinity, residency condition (urban, rural), income status (average income per month), and floor area per capita (meter) were also recorded.

Statistical analysis

NEI VFQ-25 subscales, composite score, and continuous variables were summarized by the mean and standard deviation (SD), and qualitative variables by frequency and percentages. To reduce the socioeconomic factors (occupation, education, income, residency status, and floor area per capita variables) to one composite factor used through Polychoric Principal Component Analysis method. Ordinal logistic regression was used to examine related factors such as demographic, SES, and visual function with VRQoL. All variables were evaluated by univariate analysis first, and then variables with $P < 0.25$ such as age, disease duration, education, occupation, consanguinity, SES, BCVA, color vision, and VF were candidates to enter the regression models. Pearson correlation was used to evaluate the correlation between VRQoL subscales and other variables. All tests of association were considered statistically significant at $P < 0.05$. All analyses were performed using the statistical software package STATA Statistical Software version 15.0 (StataCorp).

Results

A total of 225 patients with different types of IRDs were randomly selected from 1000 registered patients in the IRDReg® database registry. Finally, 192 individuals (response rate: 85%) were included. Among study participants, 86 (44.8%) were males. The mean ± SD of age (year), disease duration (year), and the age at symptom onset (year) of the participants were 36.62 ± 13.19, 17.97 ± 12.56, and 18.50 ± 13.94, respectively. Other demographic characteristics are illustrated in Table 1. The frequency distribution of study participants was RP in 40.62% (n = 78), cone-rod dystrophy in 14.06% (n = 27), Usher syndrome in 11.46% (n = 22),

Table 1: Baseline characteristics among the studied patients with the inherited retinal dystrophy

Factors	Frequency (%)
Age (years)	
15-45	150 (78.1)
45-60	29 (15.1)
≥60	13 (6.8)
Sex	
Male	86 (44.8)
Female	106 (55.2)
Marital status	
Single	91 (47.4)
Married	101 (52.6)
Occupation	
Unemployed	104 (49.1)
Employed	88 (45.8)
Education	
Under diploma	58 (30.7)
Diploma	45 (23.8)
Academic	86 (45.5)
Residency	
Urban	176 (91.7)
Rural	16 (8.3)
Consanguinity	
No	50 (26.0)
First cousin	95 (45.5)
Second cousin	47 (24.5)
Visual impairment	
Normal vision	45 (23.4)
Mild visual impairment	11 (5.7)
Severe visual impairment	58 (30.2)
Blind	77 (40.1)

Stargardt disease in 7.8% (n = 15), LCA in 6.78% (n = 13), cone dystrophy in 5.73% (n = 11) individuals. Furthermore, other types of IRDs such as Bestrophinopathy, Choroideremia, Gyrate dystrophy, X-Linked retinoschisis, syndromic RP, Goldmann Favre syndrome, fundus flavimaculatus, congenital stationary night blindness, central areolar dystrophy, and best disease were identified in 13.55% (n = 33) of the study participants.

Patients were also evaluated for visual function, and the results present that mean ± SD BCVA, present glasses visual acuity, and near VA of the participants were 1.28 ± 1.07, 1.29 ± 1.07, and 1.15 ± 1.18 logMAR, respectively. Sixty-six (34.55%) patients had color vision deficiency (CVD), 48 (25.13%) had a normal color vision, and 77 (40.31%) were not able to respond to the Ishihara test due to their severe vision loss (NA).

The mean ± SD of the NEI VFQ-25 composite score was obtained 45.0 ± 23.6. Twelve calculated subscales are represented in Table 2.

All subscales which were obtained from the NEI VFQ-25 questionnaire, with the exception of general health, mental health, and ocular pain had a significant negative correlation with logMAR BCVA and near VA [Table 3]. The highest significant negative correlations were between subscales of near activities (r = −0.74), distance activities (r = −0.71), social functioning (r = −0.68), and color vision (r = −0.61) with logMar BCVA.

The results of ordinal logistic regression analysis showed significant associations between VRQoL and age, occupational status, logMAR BCVA, and color vision. The odds of increasing the level of VRQoL decreases by 9% with an increase in age (per year) with an odds ratio (OR) of 0.91 (95% confidence interval [CI]: 0.87–0.94). The odds of employed patients increasing the level of VRQoL was 1.37 (95% CI: 1.05–4.74) times that of unemployed patients. For each unit decrease in score of logMAR BCVA, the odds of increasing the level of VRQoL was 69% (OR = 0.31, CI: 0.19–0.49). The odds of increasing the level of VRQoL in patients with normal color vision increased 92% (OR = 1.92, CI: 1.74–5.01), and in blind patients (NA to do Ishihara test) decreased 64% (OR = 0.64, CI: 0.25–0.87) compare to those have CVD [Table 4].

Discussion

The finding of this study showed that VRQoL among Iranian patients with IRD was low. There was a statistically significant relationship between VRQoL with age, employment, logMAR BCVA, and color vision.

The overall VRQoL score in the studied patients was 45. This score was reported to be 68.4, 52.4, 59.0, 58.1, and 52.7 among IRD patients with subtypes of the disease in Japan, Sweden, Germany, France, and South Korea, respectively.[7,16,19,25,26] The VRQoL score obtained in this study was the lowest among similar studies, which is consistent with the result of a study conducted by Azizi *et al*. on RP patients in Shiraz (47.6).[17] This difference observed in Iran with developed and European countries can be due to the low level of health, treatment, and rehabilitation status, lower SES, and diagnosis of the disease at the advanced stages in Iran. It means that the different VRQoL scores are not necessarily due to ophthalmic disorder and can depend on socioeconomic conditions, lifestyle, and other issues that are not matched in the present study.

Among VRQoL subscales in the studied patients, the lowest score was obtained for the mental health and general vision subscales. The highest score was also related to the ocular

Table 2: Vision-related quality of life among adult patients with inherited retinal dystrophy and its subtypes based on the National Eye Institute Visual Functioning Questionnaire-25

Factors	Inherited retinal dystrophy (n=192)	Retinitis pigmentosa (n=78)	Usher syndrome (n=22)	Cone rod dystrophy (n=27)	Laber's congenital amerosis (n=13)	Stargardt (n=11)	Cone dystrophy (n=11)	Others (n=32)	P (*post hoc* test)
General Health	64.5±17.3	59.2±18.7*	59.7±18.8	65.9±17.3	72.7±9.8	68.5±7.3	65.7±15.6	71.5±12.4*	0.003
General vision	36.7±22.2	32.9±23.4	36.6±21.1	34.1±14.4	36.5±24.6	48.3±23.0	31.8±20.6	43.8±24.9	0.18
Mental health	35.1±27.8	32.9±27.3	24.8±26.1‡	33.7±26.9‡	53.1±28.3	49.0±23.5‡	38.6±28.7	33.6±29.2	0.04
Ocular pain	85.6±21.8	83.5±24.3	88.1±19.5	84.7±20.0	85.6±20.9	91.7±19.3	87.5±22.4	85.9±21.5	0.90
Near activities	38.9±34.7	38.1±38.2	53.4±34.9	30.7±28.5	29.0±23.4	40.3±33.4	35.0±28.8	45.0±33.8	0.17
Distance activities	42.5±31.9	38.8±32.8	48.9±27.9	38.5±29.8	29.1±28.9	55.0±32.2	45.3±35.1	48.4±32.5	0.23
Peripheral vision	45.8±38.2	34.4±34.4†	27.3±24.3‡,†	56.4±37.7	33.6±28.8*	81.7±32.0*,‡,†	52.3±41.0	63.3±39.1‡	<0.001
Color vision	51.0±39.2	50.0±42.4	42.5±35.1	43.5±37.1	43.8±42.3	61.7±36.4	43.2±37.2	53.1±36.3	0.52
Social functioning	49.6±37.5	46.5±38.7	56.1±32.7	49.1±34.5	39.7±37.8	57.2±36.8	56.1±32.7	53.9±38.6	0.78
Role difficulties	42.9±27.4	37.0±27.5	40.6±30.6	44.0±25.6	47.1±22.6	55.8±25.9	55.1±26.8	44.9±27.2	0.14
Dependency	54.3±33.0	48.2±33.0▽	45.7±34.0§	60.6±30.1	57.7±28.9	83.1±23.0▽§	65.9±26.0	50.0±34.6	0.003
Composite Score	45.0±24.3	41.6±25.1	46.2±22.8	43.9±22.1	45.5±21.5	56.9±22.9	48.2±24.4	47.9±26.3	0.40

‡‡ The mean difference between these three groups is significant using the Scheffe post hoc test. *The mean difference between these two groups is significant using Scheffe post hoc test. §The mean difference between the two groups is significant using the Scheffe post hoc test. ▽The mean difference between the two groups is significant using the Scheffe post hoc test. All values reported in the table are mean±SD. SD: Standard deviation

pain and general health subscales, which is consistent with the results of other studies.[16,19,25-28] Considering that in all similar studies, the lowest score belonged to the mental health subscale, it can be concluded that IRDs had the most destructive effect on patients' mental health.

Table 3: Correlation between vision-related quality of life and logMAR best corrected visual acuity and near visual acuity among the adult patients with inherited retinal dystrophy (*n*=192)

	BCVA		Near VA	
	r	*P*	*r*	*P*
Subscales				
General health	−0.06	0.42	−0.08	0.35
General vision	−0.60	<0.001	−0.63	<0.001
Mental health	−0.03	0.65	−0.06	0.44
Ocular pain	−0.11	0.12	−0.14	0.07
Near activities	−0.74	<0.001	−0.74	<0.001
Distance activities	−0.71	<0.001	−0.70	<0.001
Peripheral vision	−0.40	<0.001	−0.41	<0.001
Color vision	−0.61	<0.001	−0.62	<0.001
Social functioning	−0.68	<0.001	−0.68	<0.001
Role difficulties	−0.32	<0.001	−0.31	<0.001
Dependency	−0.40	<0.001	−0.39	<0.001
Composite score	−0.64	<0.001	−0.64	<0.001

VA: Visual acuity, BCVA: Best corrected VA

The mean logMAR BCVA score is 1.28 in the present study, which is higher than the same score obtained in other studies in Greece (0.74), France (0.63), Japan (0.66), and South Korea (0.63),[7,25,28,29] which in turn indicates that Iranian patients suffer from more severe visual impairment than those in other countries, which are often the developed ones. A previous study on RP patients in Iran reported a mean logMAR BCVA score of 1.1,[17] which is roughly similar to the present study. The lower the VA, the lower QoL will be. Moreover, according to studies, VA can somehow be a proxy of VRQoL.[7,12] Other previous studies that have examined visual function with VRQoL indices have also reported a significant relationship between VRQoL with improved VA as well as VF.[16,19,28,30-32] The present study showed no statistically significant relationship between VRQoL and VF, probably because this study VF was investigated by asking a question from the patient ("Do you have VF disorder or not?"), but other studies used the standard test of VF.

The present study showed a significant inverse relationship between age and QoL, consistent with other studies.[33-36] However, in a study, Hahm *et al*. found no significant relationship between VRQoL and age, as well as age with the onset of symptoms.[16] Considering the chronic and progressive nature of this disease and the gradual visual loss, VA decreases with increasing age, and such decrease will, in turn, affects a person's performance and daily activities

Table 4: Results of ordinal logistic regression to assess the sociodemographic characteristics and visual function with vision-related quality of life among the participants (*n*=192)

Variables	Vision-related QoL				OR (95% CI)
	0-25	25-49	50-75	75-100	
Age (years), mean±SD	44.2±15.8	38.9±10.8	32.4±10.9	30.8±13.5	0.91 (0.87, 0.94)
Education (frequency)					
Under diploma	22	17	11	8	1.00
Diploma	10	12	16	9	1.79 (0.68, 4.71)
Academic	4	28	31	14	1.99 (0.84, 4.74)
Occupation (frequency)					
Unemployed	28	35	23	18	1.00
Employed	8	32	35	13	1.37 (1.05, 4.74)
Consaguinity (frequency)					
No	9	18	19	4	1.00
First cousin	20	33	27	15	1.74 (0.79, 3.79)
Second cousin	7	16	12	12	3.45 (0.32, 8.98)
Duration of illness (year), mean±SD	25.4±15.8	18.6±12.5	16.4±9.7	10.4±8.0	1.03 (0.99, 1.06)
Socioeconomic status (mean±SD)	1.02±0.38	1.37±0.45	1.39±0.44	1.31±0.44	1.70 (0.87, 3.27)
LogMAR BCVA (mean±SD)	2.4±0.8	1.5±1.1	0.7±0.6	0.4±0.4	0.31 (0.19, 0.49)
Color vision					
Normal	1	9	20	18	1.92 (1.74, 5.01)
NA	31	34	10	2	0.64 (0.25, 0.87)
Defect	17	21	19	9	1.00
Vision field					
Normal	5	11	12	11	1.00
Disorder	23	43	36	13	0.76 (0.33, 1.77)

All variables entered together into the model (backward stepwise method). NA: Not applicable, SD: Standard deviation, QoL: Quality of life, CI: Confidence interval, OR: Odds ratio

and lead to several problems such as inability to read and write, loss of job, and dependence on others to walk. Furthermore, ultimately leading to a decrease in QoL in these patients. Moreover, considering the positive and significant relationship between the age of onset of symptoms and QoL, this relationship can be explained by the fact that the higher the age of onset of symptoms, the fewer years lived with visual impairment will be; therefore, it will have less impact on a person's QoL.

The results of this study showed that QoL is affected by modifiable and nonmodifiable factors such as age, duration of disease, and VA. Considering most of the determinants of QoL of IRD patients are nonmodifiable, health managers and policymakers are advised to enhance the welfare and QoL of these patients by considering the modifiable variables such as employment, level of education, SES, access to insurance services, placing these patients in the classification of special patients, providing welfare facilities in urban service centers and public environments such as the installation of fluorescence on stairs, sidewalks, and special passages (given that most patients suffer from night blindness, and commuting from the stairs as well as night walking is a problem for most of these patients, it is important to create employment opportunities, increase the number of associations, and allow patients to participate in meetings where other patients with the same disease are present to improve their morale, and to allocate funds for visual aids, access to services). In addition to the aforementioned, age is regarded as an important variable in the incidence of disease complications; therefore, the codification of rules on genetic counseling for couples, if possible, and screening and early detection of disease cases can be used as a preventive measure in the primary and secondary preventions, respectively. However, considering the current situation and absence of an accurate and efficient system for timely diagnosis and treatment, using symptomatic therapies and rehabilitation services in the form of tertiary prevention can also be beneficial.

In conclusion, this study shows an overview of the current VRQoL of IRD patients with suggestions to other researchers on other types of QoL, especially in the mental dimension. It is also recommended to carry out further intervention studies on the impact of factors such as counseling, behavioral therapy, coping strategies, strategies to improve VA, and the impact of exercise on VRQoL of these patients. In addition, these patients need psychological counseling to adapt themselves to the disease and new skills training to bring them back to normal life.

Financial support and sponsorship

This research was supported by Shahid Beheshti University of Medical Sciences.

Conflicts of interest

There are no conflicts of interest.

References

1. Coco-Martin RM, Diego-Alonso M, Orduz-Montaña WA, Sanabria MR, Sanchez-Tocino H. Descriptive study of a 488 patients cohort with inherited retinal diseases and a low genetic diagnosis rate. Research Square 2020:2-19.
2. Duncan JL, Pierce EA, Laster AM, Daiger SP, Birch DG, Ash JD, *et al.* Inherited retinal degenerations: Current landscape and knowledge gaps. Transl Vis Sci Technol 2018;7:6.
3. Motta FL, Martin RP, Filippelli-Silva R, Salles MV, Sallum JM. Relative frequency of inherited retinal dystrophies in Brazil. Sci Rep 2018;8:15939.
4. Bertelsen M, Jensen H, Bregnhøj JF, Rosenberg T. Prevalence of generalized retinal dystrophy in Denmark. Ophthalmic Epidemiol 2014;21:217-23.
5. Garip G, Kamal A. Systematic review and meta-synthesis of coping with retinitis pigmentosa: Implications for improving quality of life. BMC Ophthalmol 2019;19:181.
6. Koenekoop RK. An overview of Leber congenital amaurosis: A model to understand human retinal development. Surv Ophthalmol 2004;49:379-98.
7. Chaumet-Riffaud AE, Chaumet-Riffaud P, Cariou A, Devisme C, Audo I, Sahel JA, *et al.* Impact of retinitis pigmentosa on quality of life, mental health, and employment among young adults. Am J Ophthalmol 2017;177:169-74.
8. Bakkar MM, Alzghoul EA, Haddad MF. Clinical characteristics and causes of visual impairment in a low vision clinic in northern Jordan. Clin Ophthalmol 2018;12:631-7.
9. Levinson JD, Joseph E, Ward LA, Nocera JR, Pardue MT, Bruce BB, *et al.* Physical activity and quality of life in retinitis pigmentosa. J Ophthalmol 2017;2017:6950642.
10. Angeles-Han ST, Griffin KW, Harrison MJ, Lehman TJ, Leong T, Robb RR, *et al.* Development of a vision-related quality of life instrument for children ages 8-18 years for use in juvenile idiopathic arthritis-associated uveitis. Arthritis Care Res (Hoboken) 2011;63:1254-61.
11. Sugawara T, Sato E, Baba T, Hagiwara A, Tawada A, Yamamoto S. Relationship between vision-related quality of life and microperimetry-determined macular sensitivity in patients with retinitis pigmentosa. Jpn J Ophthalmol 2011;55:643-6.
12. Mangione CM, Lee PP, Pitts J, Gutierrez P, Berry S, Hays RD. Psychometric properties of the National Eye Institute Visual Function Questionnaire (NEI-VFQ). NEI-VFQ Field Test Investigators. Arch Ophthalmol 1998;116:1496-504.
13. Coleman AL. Development of the 25-item national eye institute visual function questionnaire. Evid Based Ophthalmol 2002;3:58-9.
14. Deramo VA, Cox TA, Syed AB, Lee PP, Fekrat S. Vision-related quality of life in people with central retinal vein occlusion using the 25-item National Eye Institute Visual Function Questionnaire. Arch Ophthalmol 2003;121:1297-302.
15. Varma R, Wu J, Chong K, Azen SP, Hays RD, Los Angeles Latino Eye Study Group. Impact of severity and bilaterality of visual impairment on health-related quality of life. Ophthalmology 2006;113:1846-53.
16. Hahm BJ, Shin YW, Shim EJ, Jeon HJ, Seo JM, Chung H, *et al.* Depression and the vision-related quality of life in patients with retinitis pigmentosa. Br J Ophthalmol 2008;92:650-4.
17. Azizi F, Riazi A, Norouzzadeh H, Tabatabaee SM. Quality of life in patients with retinitis pigmentosa in Shiraz. J Rehab Med. 2018;6:168-74.
18. Kay C, Williamson N, Bradley H, Barclay M, Sims J, Arbuckle R, *et al.* Qualitative interviews with patients and caregivers regarding visual function impairments and impacts on vision-dependent activities of daily living and health-related quality of life in RPE65-related retinitis pigmentosa and leber congenital amaurosis. Invest Ophthalmol Vis Sci 2021;62:3589.
19. Prokofyeva E, Bernd A, Wilke R, Zrenner E. Visual-related quality of life in patients with inherited retinal dystrophies – A baseline for assessing clinical trial efficacy. Acta Ophthalmol 2010;88:445-54.
20. Sabbaghi H, Daftarian N, Suri F, Mirrahimi M, Madani S, Sheikhtaheri A, *et al.* The first inherited retinal disease registry in Iran: Research protocol and results of a pilot study. Arch Iran Med 2020;23:445-54.
21. Asgari S, Hashemi H, Nedjat S, Shahnazi A, Fotouhi A. Persian version of the 25-item National Eye Institute Visual Functioning

Questionnaire (NEI-VFQ 39): A validation study. Iran J Ophthalmol 2011;23:5-14.
22. Brämer GR. International statistical classification of diseases and related health problems. Tenth revision. World Health Stat Q 1988;41:32-6.
23. World Health Organization. World Report on Vision. Geneva: World Health Organization; 2019.
24. Eskridge JB, Amos JF, Bartlett JD. Clinical Procedures in Optometry. United States: Lippincott Williams & Wilkins; 1991.
25. Seo JH, Yu HG, Lee BJ. Assessment of functional vision score and vision-specific quality of life in individuals with retinitis pigmentosa. Korean J Ophthalmol 2009;23:164-8.
26. Burstedt MS, Mönestam E. Self-reported quality of life in patients with retinitis pigmentosa and maculopathy of Bothnia type. Clin Ophthalmol 2010;4:147-54.
27. Hassan-Karimi H, Jafarzadehpur E, Blouri B, Hashemi H, Sadeghi AZ, Mirzajani A. Frequency domain electroretinography in retinitis pigmentosa versus normal eyes. J Ophthalmic Vis Res 2012;7:34-8.
28. Sumi I, Matsumoto S, Okajima O, Shirato S. The relationship between visual disability and visual scores in patients with retinitis pigmentosa. Jpn J Ophthalmol 2000;44:82-7.
29. Yioti G, Stefaniotou M, Ziavrou I, Kotsis K, Hyphantis T. Illness perceptions, psychiatric manifestations, and quality of life in patients with inherited retinal dystrophies. Semin Ophthalmol 2017;32:428-37.
30. Frost A, Eachus J, Sparrow J, Peters TJ, Hopper C, Davey-Smith G, *et al.* Vision-related quality of life impairment in an elderly UK population: Associations with age, sex, social class and material deprivation. Eye (Lond) 2001;15:739-44.
31. Ctori I, Ahmad S, Subramanian A, Oskis A. Associations between adult attachment and vision-related quality of life in visually impaired individuals. Psychol Health Med 2021;26:940-6.
32. Sugawara T, Hagiwara A, Hiramatsu A, Ogata K, Mitamura Y, Yamamoto S. Relationship between peripheral visual field loss and vision-related quality of life in patients with retinitis pigmentosa. Eye (Lond) 2010;24:535-9.
33. Yibekal BT, Alemu DS, Anbesse DH, Alemayehu AM, Alimaw YA. Vision-related quality of life among adult patients with visual impairment at university of Gondar, Northwest Ethiopia. J Ophthalmol 2020;2020:9056097.
34. Adigun K, Oluleye TS, Ladipo MM, Olowookere SA. Quality of life in patients with visual impairment in Ibadan: A clinical study in primary care. J Multidiscip Healthc 2014;7:173-8.
35. Nirmalan PK, Tielsch JM, Katz J, Thulasiraj RD, Krishnadas R, Ramakrishnan R, *et al.* Relationship between vision impairment and eye disease to vision-specific quality of life and function in rural India: The Aravind Comprehensive Eye Survey. Invest Ophthalmol Vis Sci 2005;46:2308-12.
36. du Toit R, Palagyi A, Ramke J, Brian G, Lamoureux EL. The impact of reduced distance and near vision on the quality of life of adults in Timor-Leste. Ophthalmology 2010;117:2308-14.