

Optometry

Journal of the American Optometric Association



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Stargardt's macular dystrophy— a patient's perspective

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Purpose: The purpose of this paper was to evaluate life experience, including disease characteristics, daily-living activities, employment, interactions with health care professionals, and support services in patients diagnosed with Stargardt's macular dystrophy.

Methods: More than 200 patients with Stargardt's disease responded to a 68-question survey. Results were analyzed using SAS Statistical Analysis Package.

Results: Of 203 responders, 142 (70%) were women. Early disease onset occurred in more than 60% of patients. Blurred vision (134, 66%) and glare (183, 90.1%) were the leading symptoms reported. Reading (116, 57.1%), driving (86, 42.4%), and recognizing faces (66, 32.5%) were daily-living activities most difficult to perform. Patients with early disease onset had worse vision at presentation ($p = 0.001$), faster progression of visual loss ($p = 0.007$), and were more often diagnosed with a non-physiological visual loss ($p = 0.007$). Patients with late disease onset had more difficulty with orientation and coping skills ($p = 0.02$). Sixty-five percent of evaluated adults (108 of 165) were employed.

Conclusions: Although the study illustrates that patients with Stargardt's disease can contribute and function well in contemporary society, issues related to depression, and availability and quality of health care, are still major concerns for this patient population. The study shows differences in progression of visual loss between patients, with early versus late disease onset indicating that age at onset and visual acuity at presentation might be two important factors influencing patient's visual prognosis. Finally, the study suggests parallels in psychological profiles between late age at onset Stargardt's disease and age-related macular degeneration patients.

Key Words: Age-related macular degeneration, Stargardt's disease

Stargardt's macular dystrophy is considered to be one of the most common macular dystrophies, accounting for approximately 7% of hereditary retinal degenerations.¹ The majority of patients begin manifesting their symptoms in childhood, with a slow progression of bilateral visual loss over a period of several decades. Due to clinical characteristics of the disease, affected individuals are required to cope with a disability of legal blindness throughout the majority of their life.^{2,3} Since Stargardt's macular degeneration is not a prevalent disease, the literature has not yet addressed many issues pertinent to this patient population and socioeconomic data are lacking.

The visual data collection of a large patient population is usually associated with individual testing with the use of technical equipment and highly trained personnel in a medical office setting. Unfortunately, obtaining reliable data pertinent to the prevalence and epidemiological characteristics of a rare genetic disease, using standard laboratory and clinical measures, is rather difficult—if not impossible—due to staffing, technical, geographic, and financial limitations. Over the past decade, a series of studies have been conducted that proved the hypothesis that a self-report inventory is an acceptable and reliable format for obtaining information from carefully selected, large groups of patients.⁴⁻¹⁰

We applied the self-report method to a population with Stargardt's macular degeneration and designed a disease-specific questionnaire to evaluate the perceived impact of this disease on patients' live experience. We compared visual function levels, age at disease onset, a family history of the disease and disease duration with their effect on disease

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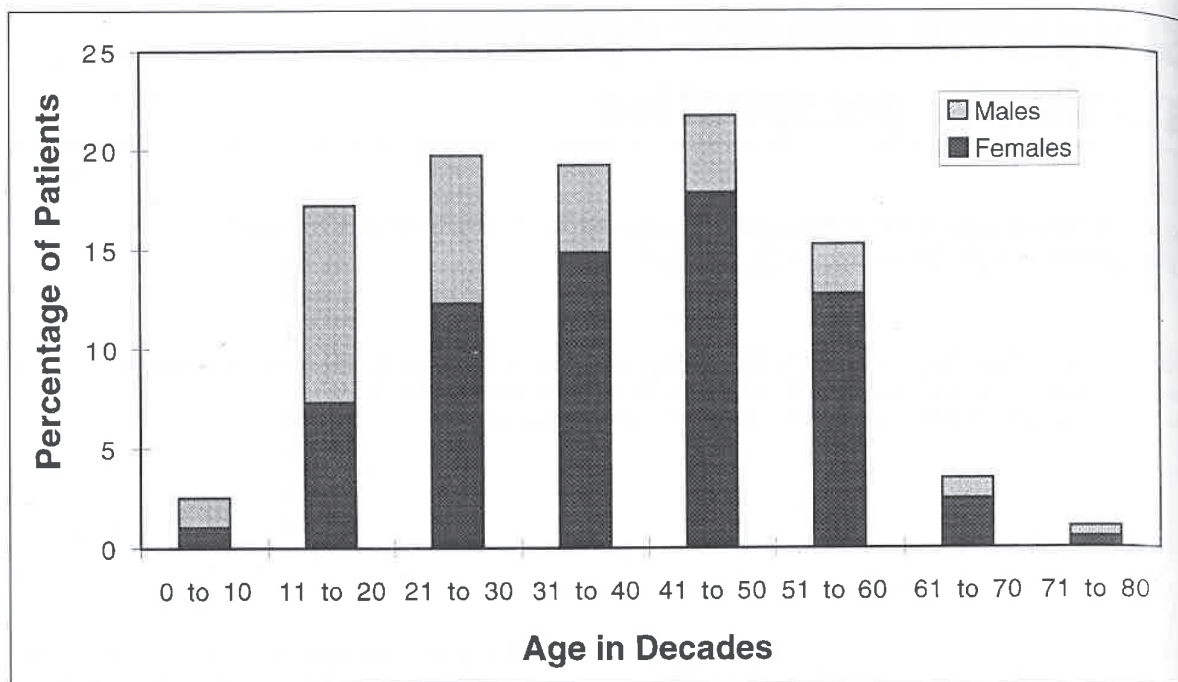


Figure 1 Age and gender distribution in the study population.

characteristics, performance of tasks of daily living, employment, use of visual aids, emotional status, and interaction with health care professionals and support organizations.

Methods

A 68-question survey was developed in our laboratory. Its format was based on a number of previously developed questionnaires used in geriatric (i.e., the *Vision-Specific Sickness Impact Profile*,⁸ the *Community Disability Scale*,¹¹ the *Geriatric Depression Scale*,¹¹ the *General Health Questionnaire*⁸) and low-vision patient populations (i.e., the *Visual Activity Questionnaire*,¹⁰ the *National Institute of Health Visual Function Questionnaire*,⁷ the *Visual Status Inventory*⁴) as described in the recent literature. We also sought a direct input from patients affected by the disease in regard to issues pertinent to them in daily-living situations. The large-print questionnaire was distributed to 1,000 members of Macular Degeneration International Inc. Inclusion criteria for the study consisted of: a known diagnosis of Stargardt's macular degeneration, confirmed by a vision care professional (an optometrist or an ophthalmologist) and the ability to complete and return the questionnaire. The waiting period for responses to the study mailing was 3 months. Of 277

patients who responded to the questionnaire, 203 carried the diagnosis of Stargardt's macular dystrophy. The remaining 74 patients had other macular diagnoses and were excluded. Questionnaires were either answered by the adult patients themselves, or by parents of minors with the disease (8 to 14 years of age group).

The survey was divided into two parts: Part I included general demographic and disease-related questions; Part II addressed visual and physical function and was subdivided into: central vision, general orientation, and mobility sections. On receiving all of the responses to survey questions, it occurred that 10 of 68 questions were answered by less than 50% of participants and an additional 4 questions were as frequently misunderstood. These 14 questions were excluded from the statistical analysis.⁵ Therefore, the final number of questionnaire items reviewed was reduced to 54—35 questions in part I and 19 questions in part II (see Appendix).

Patients were grouped on the basis of presence or absence of a family history of the disease, age at onset (early onset by 15 years of age *versus* 16 to 29 years of age *versus* late onset at or after 30 years of age), visual acuity level (vision better than

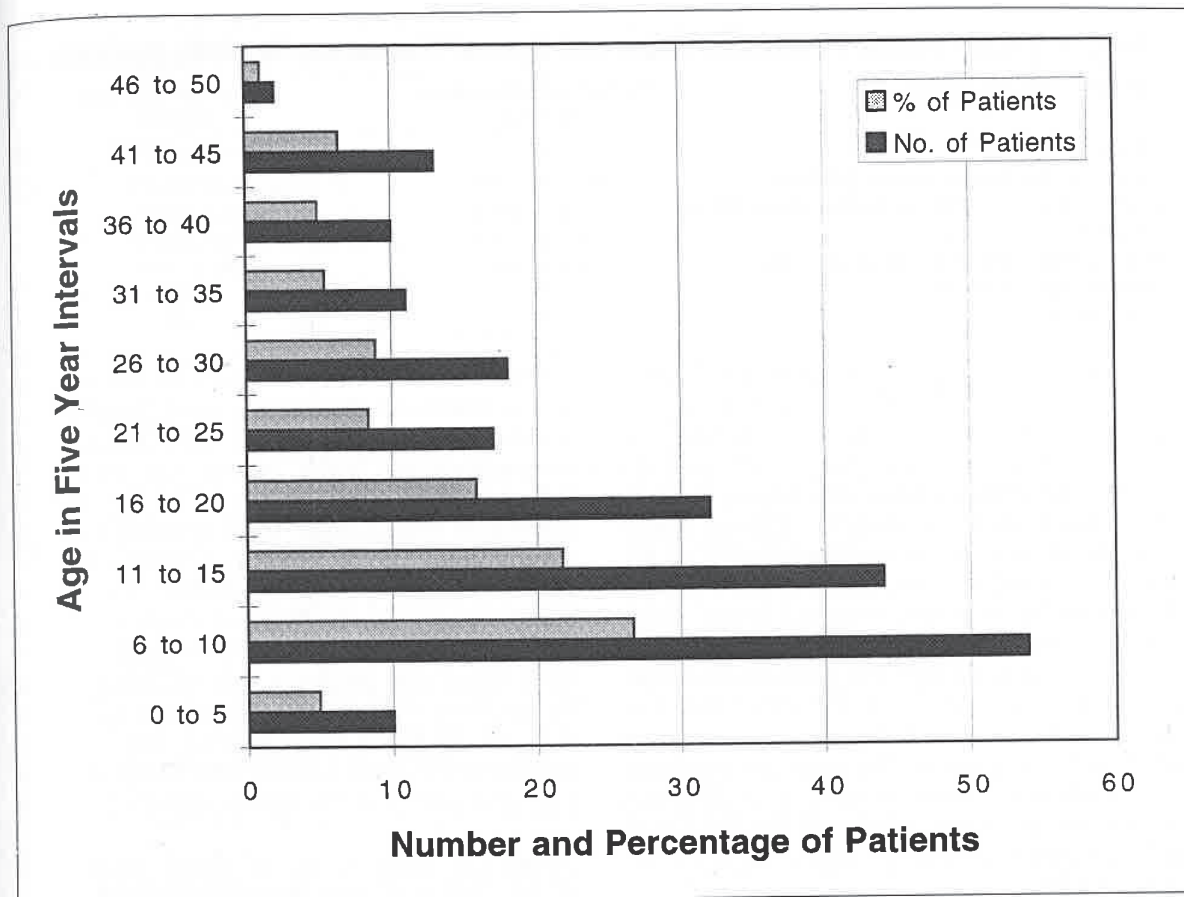


Figure 2 Age at onset of disease in the study population.

20/200 at the time of the survey *versus* 20/200 and worse), and duration of disease (up to 10 years, 11 to 20 years, 21 to 30 years, 31 years or longer). Functional, socioeconomic, and clinical characteristics of these groups were compared using the SAS Statistical Analysis Package,¹² the Chi-square test for independence and the Mantel-Haenszel test for trend.¹³ The questionnaire's internal consistency or degree of correlation among items was evaluated by percentage of agreement between specific sets of questions (data not shown).

Results

Please note that many steps of the statistical analysis presented in this article were performed on slightly smaller groups of patients than total quoted numbers of participants, depending on how many of the study patients answered all of the questions pertaining to a given analyzed topic.

Part I—General Data

Two hundred three persons with Stargardt's macular dystrophy responded to the survey. Approximately seventy percent (142) of responders were women. Ages of responders ranged from 8 to 76 years, with the mean of 35.7 (*see* Figure 1). Early age at disease onset (1st or 2nd decade of life) was reported by more than 60% of the study population (*see* Figure 2). Eighty-one patients (39.9%) reported vision equal to or better than 20/100; two reported vision of 20/150; 49 (24.1%) reported vision of 20/200; and 13 (6.4%) reported vision worse than 20/200 at the time of initial diagnosis. Only 24 patients (11.8%) maintained vision of 20/100 or better at the time of the survey (*see* Table 1). The first symptoms noticed by patients included: blurred vision, difficulty with school work (not seeing the blackboard), sitting close to the television or reading materials, and driving problems. Twenty individuals (10%) were identified at routine eye screenings. One hundred

Table 1. Visual acuity at disease onset and time of the survey in study patients

Visual acuity level	Onset of disease n (%)	Time of survey n (%)
20/40 or better	39 (19.2%)	7 (3.4%)
Worse than 20/40, up to 20/100	42 (20.7%)	17 (8.4%)
Worse than 20/100 to better than 20/200	2 (0.9%)	1 (0.5%)
20/200	49 (24.1%)	75 (36.9%)
Worse than 20/200, up to 20/400	13 (6.4%)	55 (27.1%)
Worse than 20/400	0	13 (6.4%)
Unknown	58	35

fourteen patients (56.2%) had abnormal color vision and 130 (64%) admitted to problems with adaptation to darkness. A vast majority of patients experienced glare (183, 90.1%) and 120 (59.1%) preferred to wear sunglasses indoors. When asked about comfortable lighting conditions, almost equal amounts of patients (97, 47.8% *versus* 91, 44.8%) selected bright *versus* dim light but a majority of people reported better vision indoors (109, 53.7%) in comparison to outdoors (56, 27.6%). Over seventy five percent of the evaluated population (153 patients) stated that it is easier for them to read dark print on a white paper than the opposite. The most difficult tasks to cope with or perform at the time of diagnosis and the survey were reading and driving. Other daily-living problems varied depending upon disease duration.

More than 50% of evaluated individuals (104 patients) were diagnosed within the first year of the onset of symptoms. One hundred thirty-three patients (more than 65%) were diagnosed within the first three visits. The majority of individuals (119, 58.6%) were diagnosed in a private practice setting. Retina specialists made the diagnosis in 100 patients (49.3%), while 85 (41.9%) were seen by general ophthalmologists and 11 (5.4%) were initially examined by optometrists. Forty-six patients (22.7%) were originally diagnosed with conditions other than Stargardt's disease, 47 (23.2%) were not given any diagnosis, and 18 (8.9%) were told that they have a nonphysiological visual loss (examination was rendered normal). Only 109 patients (53.7%) felt they were adequately counseled at the time of the diagnosis. Physicians were the source of referrals to support services in 120 of the cases (59.1%). The school system and family represented the other 10% (20 of 203), but more than 20% of patients (43 of 203) were never referred and sought help independently. Services recommended by health

care professionals included: the State Associations for the Blind and Visually Impaired (39.9%), low-vision centers (21.7%), and Macular Degeneration International Inc. (5.9%). Patients were most often followed up on yearly basis (110, 54.2%), but frequency of visits ranged from monthly to every several years. The majority of patients underwent fundus photography (184, 90.6%), fluorescein angiography (155, 76.4%), and electroretinography (109, 53.7%). More than 92% of patients (187) used visual aids (both for near or distance) and the majority were happy with their performance (173, 85.2% found them helpful), but only 134 patients (66%) were content with the availability, cost, and quality of low-vision services provided.

Adults (21 years of age or older) represented 81.3% (165 of 203) of the study population and 65% of this subgroup (108 of 165) were employed. Professions in the health care industry predominated (23, 21.3%), followed by business (19, 17.6%), teaching (10, 9.3%), management (10, 9.3%), and clerical jobs (9, 8.3%).

Issues related to depression were raised by more than half of the patients responding to the survey (105, 51.7%) and 93.3% of individuals from this subgroup (98 of 105) stated their feelings were related to visual loss.

Part II—Subgroup Analysis

A. Family history

One hundred twenty-eight individuals (63.1%) had no family history of Stargardt's macular dystrophy. Comparison of subgroups of patients—with and without a family history of the disease—revealed that both groups had very similar disease characteristics and experience with health care services. A lack of a family history of the disease did not significantly delay a correct diagnosis ($p = 0.116$), but this subgroup was

more frequently diagnosed with a nonphysiological visual loss (13, 10.2% versus 1, 1.4%, respectively).

B. Age at disease onset

One hundred eight patients (53.2%) had a disease onset within the first 15 years of their life; 63 (31%) between the ages of 16 and 29 years; and 32 (15.8%) at or after 30 years of age. A mean age of patients in the first group was 14.2 years ($SD \pm 2.9$) versus 35.1 ($SD \pm 8.6$), and 57.6 ($SD \pm 6.1$) in the second and third groups, respectively.

Patients in the early onset group (15 years of age and younger) manifested worse levels of visual acuity (47 of 81 patients from the early onset group had vision of 20/200 or worse versus 17 of 64 patients with the age of disease onset after 16 years of age, $p = 0.001$) and were more often misdiagnosed (nonphysiological visual loss diagnosis in 14 of 108, 13% of cases versus 1 of 95, 1.1% in the other two groups combined, $p = 0.007$). Patients with early age of onset usually reported problems at school (46, 42.5%), were noticed to sit close to television or reading material (87, 80.6%), or were picked up during routine eye screenings (17 of 108, 15.7% of patients in the early onset group versus 3 of 95, 3.3% in the other groups combined, $p = 0.02$).

Patients in the late onset group (30 years of age and older) were seen more frequently in followup (12 of 32, 37.5% in the late onset group versus 36 of 171, 21% in the other two groups combined were seen every 6 months) and were more often holders of a valid driver's license (16 of 32, 50% in the late onset group versus 14 of 71, 19.7% in the early onset group, $p = 0.0001$; 37 patients in the early onset group were minors). They also admitted to much higher level of difficulty with coping with the diagnosis and visual prognosis of the disease than patients in the early onset group (13 of 32, 40.6% and 25 of 63, 39.7% in both late onset groups versus 23 of 108, 21.2% in the early onset group, $p = 0.02$).

The age of disease onset did not seem to influence patients' ability to perform in the job market (46 of 71, 64.8% versus 42, 66.7% versus 22, 71% were employed in each group, respectively; 37 patients in the early onset group were minors).

C. Visual acuity

Twenty five patients had vision better than 20/200 at the time of the survey, in comparison with 143 patients whose vision was 20/200 or worse (information relating to vision at disease onset is quoted under general data heading).

Patients in the better visual acuity group (at the time of survey) had better vision at presentation (12 of 25, 48% had visual acuity of 20/50 or better at presentation versus 24 of 143, 16.7% of patients in the poor visual acuity group, $p = 0.001$); a later onset of the disease (12 of 25, 48% had the disease onset in the first or second decade of life versus 105 of 143, 73.4% of patients in the poor visual acuity group, $p = 0.02$); and a shorter disease duration (18 of 25, 72% were diagnosed with the disease for less than 10 years versus 68 of 143, 47.5% of patients in the poor visual acuity group, $p = 0.04$). They were more often disturbed by the diagnosis and the visual prognosis of the disease in comparison with the poor vision group (7 of 25, 28% versus 9 of 143, 6.3%, $p < 0.0001$).

Individuals with vision of 20/200 or worse were more often diagnosed by a general ophthalmologist (69 of 143, 48.3%) while the better vision group usually had to be evaluated by a retina specialist (18 of 25, 72%; $p = 0.008$). More than 17% of patients in the poor vision group had valid driver's licenses (25 of 143) and only approximately one third (7 of 25) had restricted driving privileges.

Analysis of visual acuity levels at both presentation and time of the survey in relation to age at onset of disease revealed that individuals with an early age at onset had more profound visual loss at presentation ($p = 0.001$). The similar comparison of visual acuity levels with patients' ages proved not to be statistically significant ($p = 0.092$). Analysis of visual acuity levels at both presentation and time of the survey in relation to patient's age and age at onset of disease—controlling for disease duration—revealed that younger patients who experienced Stargardt's disease for the same length of time as older individuals experienced somewhat faster visual loss, showing that age at onset and not patient's age is a variable associated with individuals' ultimate level of vision. Overall, the majority of patients reached the level of 20/200 or worse vision dur-

Table 2. Visual acuity levels at time of survey in relation to age of disease onset controlling for disease duration in the study population*

Visual acuity level	Age at onset of disease			P value
	0 to 15 yrs n (%)	16 to 29 yrs n (%)	30 yrs and older n (%)	
Duration of disease: 0 to 10 years				
Better than 20/200	5 (14.3%)	5 (38.5%)	4 (57.1%)	0.007
20/200 or worse	30 (85.7%)	8 (61.5%)	3 (42.9%)	
Total	35	13	7	
Duration of disease: 11 to 20 years				
Better than 20/200	1 (4.4%)	2 (11.8%)	2 (18.2%)	
20/200 or worse	22 (95.6%)	15 (88.2%)	9 (81.8%)	
Total	23	17	11	
Duration of disease: 21 to 30 years				
Better than 20/200	2 (16.7%)	0	0	
20/200 or worse	10 (83.3%)	10 (100%)	5 (100%)	
Total	12	10	5	
Duration of disease: 31 years and longer				
Better than 20/200	1 (4%)	0	1 (100%)	
20/200 or worse	24 (96%)	8 (100%)	0	
Total	25	8	1	

* Subtotals of patients shown in this Table are smaller than the ones quoted in the text regarding vision, disease duration, or age at onset of disease. Not all of the survey respondents provided all of the necessary information required for the above analysis.

ing the first 10 to twenty years of the disease (see Table 2).

Duration of disease

Sixty-eight patients (33.8%) had the disease for up to 10 years; 60 (29.9%) had it for 11 to 20 years; 33 (16.4%) had it for 21 to 30 years; and 40 (19.9%) had the disease for 31 years or longer (data on two patients were unavailable). Forty-one of 51 patients (74.6%) who carried the diagnosis of Stargardt's macular dystrophy for up to 10 years were legally blind at the time of the survey, in the comparison with 103 of 112 patients (92%) who had the disease for more than 10 years ($p = 0.007$; visual acuity at the time of survey was not known in a total of 35 patients). Individuals who experienced the disease longer were seen in followup less frequently: 23 of 59 patients (39.9%) who had the disease for up to 10 years versus 24 of 119 patients (20%) who had it for more than 10 years were seen every 6 months ($p = 0.0001$). Patients who had the disease longer than 31 years (11 of 39 patients, 28.2%) reported the most difficulty finding means of transportation, while the

up-to-10 years duration group found it to be of much less concern (4 of 64 patients, 6.3%; $p = 0.02$).

Approximately half of the patients whose disease duration did not exceed 10 years could not see faces at a distance (33 of 68, 48.5%) or read street signs at night (39 of 68, 58.2%). By the time the disease had lasted for more than 30 years, almost 80% of patients could not see street signs (31 of 40, 77.5%) nor see faces from afar (32 of 40, 80%). Approximately 10% of patients who had the disease for up to 10 years could not see well at near (5 of 68, 7.5% could not read a newspaper and 8 of 68, 11.8% could not read directions on medicine bottles), in comparison with almost 30% of individuals affected by Stargardt's macular degeneration for more than 30 years who could not perform the same activities with or without appropriate visual aids (9 of 39, 23.1% could not read a newspaper and 11 of 39, 28.2% could not read directions on medicine bottles) (Note: presbyopia was not a factor, since the disease developed in early childhood among patients with the

Table 3. Statistically significant responses to orientation section questions in study population

Orientation task	Age at onset of disease			P value
	0 to 15 yrs (%)	16 to 29 yrs (%)	30 yrs and older (%)	
Can you:				
<i>Get around in unfamiliar places</i>				
No	8.41%	8.33%	12.9%	<i>p</i> = 0.06
Yes, with visual aids	25.2%	30%	41.9%	
Yes, without visual aids	66.4%	61.7%	45.2%	
<i>Find a seat in a movie theatre</i>				
No	7.4%	11.3%	17.2%	<i>p</i> = 0.04
Yes, with visual aids	12%	11.3%	20.7%	
Yes, without visual aids	80.6%	77.4%	62.1%	
<i>Use an escalator</i>				
No	0	1.6%	0	<i>p</i> = 0.07
Yes, with visual aids	7.4%	9.7%	19.4%	
Yes, without visual aids	92.6%	88.7%	80.7%	

disease duration of 30 years and who were less than 40 years of age).

Part III—Visual and Physical Function

A. Mobility

One hundred seventy-six patients (86.7%) actively exercised. More than 90% percent of the evaluated population were able to climb stairs (202, 99.5%) and walk outdoors unassisted (183, 90.1%). There were no significant differences between patients with or without a family history of the disease, different age at onset of disease, visual acuity levels, or disease duration.

B. Orientation

Approximately 90% of study participants were able to function well and maintain orientation in an unfamiliar environment. The task that was consistently the most difficult to perform in all subgroups—overall 132, 65% of patients could not perform it—was reading street signs at night. It was also noted that there was a trend for patients in the late disease-onset group to have more difficulty with orientation, although this reached only borderline levels of statistical significance (*see* Table 3).

C. Central vision

Overall, more than 60% of patients were able to perform tasks that required central visual acuity (with or without use of visual aids). The most dif-

ficult task—133, 65.5% of patients were unable to perform it—proved to be recognition of faces from a distance. This finding was directly correlated with the level of visual acuity, since only four of 147 patients (2.7%) in the poor visual acuity group could perform this task without a visual aid, in comparison with five of 25, 20% of patients with better than 20/200 vision ($p = 0.02$). Other activities directly correlated with visual acuity were reading newspaper print ($p = 0.001$), directions on medicine bottles ($p = 0.001$), or ingredients on cans of food ($p = 0.001$).

Discussion

Although self-report questionnaires have limitations and patients' responses can be influenced by nonvisual personality traits (response to stress, adaptability, independence, incentives, and motivations) if used on a large patient population, these variables become diffused across the data set, enabling observation of trends.⁶ Also, the disease-specific survey as a measure of quality of life or health status (assessing specific symptoms or areas of disability) is known to have an excellent content validity and can be used as a psychometric tool for the assessment of patients' perception of disability.^{14,15}

In our survey, disease characteristics such as age at onset, visual acuity, manifesting symptoms, etc.

were consistent with previous reports using performance-based measures (i.e., Snellen Visual Acuity, Fluorescein Angiography, Visual Field testing etc.).^{2,3}

As expected, main problem areas reported by the evaluated patient population were reading small print, driving, recognition of objects at a distance, and performance of detail-oriented activities (typing and computer work). The patients with the early disease onset had much worse vision at presentation and more rapid visual loss. These findings suggest that not only age at onset of disease, but also visual acuity at presentation may be factors that determine patients' visual prognosis.

We also found that distance vision was more severely affected in comparison with near vision in patients with Stargardt's disease, but the progression of visual loss was similar for these two categories.

Finally, we discovered that patients with a later age at onset of disease had more difficulty with both orientation skills and psychologically coping with the disease. As Marron et al.¹⁶ and Warren et al.¹⁷ reported, the orientation-mobility performance depends not only on residual vision, but also on the age at onset of visual impairment, posture and balance, intelligence, body image, auditory-tactile abilities, and personality. Since mobility performance did not differ in our patient population between different ages at onset of disease, levels of vision, or age groups, it would appear that orientation difficulties might be associated with emotional status and adjustment problems in patients with the late disease onset.

The issue of depression was raised quite often (approximately 50%) by this patient population. Groups that seemed to associate the depression with visual loss the most were patients who either had relatively good vision at the time of disease diagnosis or who were diagnosed later in life (third decade and up). The depression associated with the diagnosis of Stargardt's disease appeared to decrease with disease duration (see Table 3). This finding might indicate that depression is not a direct result of the loss of central vision, but results from the perceived disability caused by visual loss and that, with time, motivated patients can overcome and adjust to the imposed limitations.⁹

Health and vision-related quality of life issues are vital to our society, with increasing life spans and standards of living. Issues of depression, as well as overall performance among patients with both juvenile and age-related central visual loss, directly affect their life experience. The impact of psychosocial issues on age-related macular degeneration, as well as the overall geriatric population, has been extensively discussed in the recent literature.^{9,11,18,19} On the other hand, the same topics in relation to juvenile macular degeneration patients have not been addressed in depth.

Results of our questionnaire indicate the need for more uniform ways of initial evaluation and followup of patients with Stargardt's disease. The study shows that the involvement of health professionals and support services should not end with correct diagnosis, since patients with macular degeneration are required to cope with the consequences of the disease for majority of their lives. The study also suggests some parallels in psychological profiles between the late age at onset Stargardt's disease and patient populations with age-related macular degeneration that need to be further investigated.

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Appendix

Stargardt Macular Degeneration Vision Questionnaire Part I—General Information

1. Name: _____
2. Age: _____
3. Date of birth: ____/____/____
4. Sex: Male / Female
5. If employed, your profession: _____
6. Diagnosis of the eye condition: _____
7. Age at onset of first signs of the disease: _____
8. Age at diagnosis by a physician: _____
9. First symptoms: _____
10. If known: Level of vision at the time of the diagnosis: R _____ L _____
11. If known: Your current level of vision: R _____ L _____
12. If known: Who diagnosed your eye condition:
Optometrist / Ophthalmologist / Retina Specialist
Located in: Private Practice / Teaching Hospital
13. Was your condition diagnosed with a first visit: Yes / No
If no, how many visit were needed: _____
If no, what were the other diagnosis you were given: _____
14. Age of a first low vision evaluation: _____
15. Were you referred to support services at a first visit to an eye doctor: Yes / No
If no, when were you referred: _____
Which services were you referred to: _____
Who referred you: _____
16. Were the origins of the eye condition and the prognosis made clear to you initially : Yes / No
Who counseled you: _____
17. How often were you seen by a physician after you were diagnosed:

18. Have you ever had photographs of the back of your eyes taken: Yes / No
19. Have you ever had a fluorescein angiography (dye test) of the back of your eyes: Yes / No
20. Have you ever had an ERG (test of electric activity of your retinas) done: Yes / No
21. Color vision: Normal / Abnormal
22. Hearing: Normal / Abnormal

23. Difficulty adjusting to darkness: Yes / No

If yes, age of onset: _____

24. Which light conditions are more comfortable for you :

Brightly lit room / Dimly lit room / Same

25. Under which light conditions do you see better:

A. Outdoors / Indoors

B. With light source: Above / In front / Behind / Same

C. With sunglasses: On / Off / Same

D. With lights: On / Off / Same

26. What is easier for you to read:

A. Black print on white paper

B. White print on black paper

C. Same

27. Do you have trouble with glare: Yes / No

28. Are you currently licensed to drive: Yes / No

If yes: Unrestricted / Restricted

29. Sight is better: At near / At far / Same

30. Have you noticed any conditions that changed the level of your vision: Yes / No

If yes, describe:

-bright lights: Yes / No

-medical health: Yes / No

-smoking: Yes / No

-eye photographs: Yes / No

-other: _____

31. List five things that were most difficult for you when you were first diagnosed with eye disease:

1. _____

2. _____

3. _____

4. _____

5. _____

32. List five things which are most difficult for you now:

1. _____

2. _____

3. _____

4. _____

5. _____

33. Do you use vision aids: Yes / No

A. If yes, what kind: Glasses / Magnifier / Telescope / CC TV / Other

B. Is low vision care accessible to you: Yes / No

C. Are you satisfied with services: Yes / No

34. Do you have family members with the same disease: Yes / No

35. Do you often feel sad or depressed: Yes / No / Sometimes

Part II—Activity Level Evaluation

* Word *aid* used in the text refers to help or assistive device(s)

Central Vision Assessment:

- | | |
|---|-----------------------------------|
| 1. Can you thread a needle: | Without an aid / with an aid / no |
| 2. Can you read directions on medicine bottles: | Without an aid / with an aid / no |
| 3. Can you read ingredients on cans of food: | Without an aid / with an aid / no |
| 4. Can you watch TV: | Without an aid / With an aid / no |
| 5. Can you see faces across the street: | Without an aid / with an aid / no |
| 6. Can you read newspaper print: | Without an aid / with an aid / no |
| 7. Can you use a ruler: | Without an aid / with an aid / no |
| 8. Can you eat meals with a fork and a knife: | Without an aid / with an aid / no |

General Orientation Assessment:

- | | |
|--|-----------------------------------|
| 1. Can you get around in unfamiliar surrounding: | Without an aid / with an aid / no |
| 2. Can you find a seat in a movie theatre: | Without an aid / with an aid / no |
| 3. Can you find particular items in a store: | Without an aid / with an aid / no |
| 4. Can you read street signs: | |
| During the day: | Without an aid / with an aid / no |
| At night: | Without an aid / with an aid / no |
| 5. Can you prepare a meal: | Without an aid / with an aid / no |
| 6. Can you use public transportation: | Without an aid / with an aid / no |
| 7. Can you use an escalator: | Without an aid / with an aid / no |

Mobility Assessment:

1. Do you exercise: Yes / No

If yes, list 3 most common activities: _____

- | | |
|---|-----------------------------------|
| 2. Can you perform general grooming activities: | Without an aid / with an aid / no |
| 3. Can you walk outdoors unassisted at night: | Without an aid / with an aid / no |
| 4. Can you walk down steps: | Without an aid / with an aid / no |