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Educational Levels in Patients with Stargardt Disease

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INTRODUCTION

Stargardt disease, first described in 1909 by Karl Stargardt,¹ represents the most common form of juvenile onset, hereditary macular dystrophy that leads to progressive loss of (predominantly) central visual function. It is an autosomal recessive disease, affecting approximately 1 in 8000 to 1 in 10000 people in the United States.² Visual acuity (VA) can be variable,³ most often ranging from 20/20 to 20/400 and these patients often have central visual field scotomas.⁴

Previous studies have evaluated the difficulties that these patients experience in performing daily activities such as reading and driving.^{5,6} However, to our knowledge, there are no available reports in the literature concerning the educational levels attained by patients afflicted with the disease. The aim of our study was to assess the educational levels attained by patients with Stargardt disease and clinically significant impairment of their VA.

PATIENTS AND METHODS

The medical records of all patients with the diagnosis of Stargardt disease seen by one of the authors (GAF) were reviewed. Patients with documented VA of 20/70 or worse in both eyes either prior or up to the age of 22 were included in the study. All participants who were 21 years of age or older were interviewed.

Of the 545 patients with Stargardt disease who were examined in our clinic, 109 fulfilled the inclusion criteria stated above. We were able to contact 33 subjects via either a phone interview or during a follow up eye examination. In two cases where our patients were deceased, the information regarding only the educational levels was acquired by a first degree relative after obtaining his/her consent. The purpose of the study was explained to all subjects and participants were interviewed by one of the authors (AA, AT). All patients were asked the same group of 14 questions. The information that was obtained by the questionnaire included: patient demographics, the highest level of education attained, possible future educational plans, current occupational status, the use of low vision aids during school years, and the presence of any other medical condition, including an attention deficit disorder, that could possibly interfere with education.

In cases where only a high school degree was obtained, patients were asked to report whether their visual impairment or other reasons (financial or lack of interest) were

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DISCLOSURE:

The authors (AA, AT, GAF, JM) have no financial or proprietary interest in any of the products or techniques mentioned in this article.

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responsible for not pursuing further education. Subjects were asked to subjectively rate the perception of their visual function during the period of their education (poor, fair, good), the perception of their quality of life (poor, good, excellent) and whether they had a sense of accomplishment (yes, no, unsure). Additional information was collected regarding the level of education attained by the patients' parents, the financial status of the family during the period of education, and whether any additional financial support (financial aid, scholarships, grants) was received. For the purpose of data analysis, the financial status of the family was defined as lower economic class for those households with an income of less than \$40,000, middle economic class (household income \$40,000 – \$80,000) and upper economic class (household income greater than \$80,000).

Spearman's rank correlation coefficient (ρ) was used to evaluate the relationship between: the financial status of the patient's family and the highest level of education attained; the patient's subjective perception of visual function during the period of education and the patient's subjective perception of his or her quality of life.

RESULTS

Of the 33 patients included, 15 were males and 18 females. The age range of the patients was 21 to 60 years, with a mean age of 32.7 years, ($SD \pm 11.6$ years). Twenty-four patients were Caucasian, five were African American, two were Asian and two were Hispanic. The mean age of diagnosis was 14.3 years.

An analysis of the best corrected visual acuity (BCVA) documented up to and including the age of 22 years, as measured on a Snellen projection chart, showed that 10 patients (30.3%) had visual acuity better than 20/200 in the better seeing eye. The remaining 23 patients (69.7%) were classified legally blind using the criteria of central visual acuity as 20/200 or worse in the better eye. Analytically, the BCVA of the study population ranged between 20/70–20/80 in eight subjects, 20/100–20/160 in two subjects, and 20/200–20/400 in 23 subjects.

Thirty-two of the 33 patients completed high school while one completed 7th grade. Of the 32 patients who obtained a high school degree, five (15.6%) did not pursue any further education and 27 (84.4%) proceeded for further education. For the five patients who obtained only a high school degree, two reported that the main reason for not pursuing any further studies was their visual condition and the remaining three reported that the reason was not related to vision.

Five individuals (15.6%) obtained an associate degree defined as a certificate granted upon completion of two years of education beyond high school (one additional patient was enrolled for a bachelors degree after attaining an associates degree). Fifteen individuals (46.9%) obtained a bachelors degree, defined as the successful completion of four years of full-time undergraduate study or the equivalent. Three individuals (9.4%) were enrolled in college at the time of the interview: one patient was in the first year, and two patients were in their second year. Four additional patients (12.5%) had attended but did not complete a college program. In these four patients, the discontinuation of education was related to the visual impairment in one patient, partly related to the visual impairment in one patient, and unrelated to vision in two additional patients. Seven patients (21.9%) pursued a graduate degree, with six patients acquiring a master's degree and one patient a Ph.D. degree. Table 1 summarizes these findings.

There was no statistically significant relationship between the age at which patients reached a clinically significant level of low vision (defined as 20/70 or less in their better seeing eye) and their level of education ($\rho = -0.06$, $p = 0.76$). For the subjects with only a high school

degree, the average age at which low vision was recorded was 15.5 years. For the subjects who attended college, the average age at which low vision was recorded was 15.1 years. For the subjects with education beyond a bachelor's degree, the average age at which low vision was recorded was 14.7 years. These average ages are all within one year, indicating that the age at which patients reached a clinically significant low vision was not related to their educational levels.

Two patients included had significant medical disorders which had the potential to interfere with their studies; one was affected with severe Crohn's disease and one with a bipolar psychiatric disorder. Among these two patients, one completed a masters degree while the other discontinued high-school.

Regarding the use of low vision aids during school years, 26 patients (78.8%) used low vision aids, most commonly magnifiers, and/or a closed circuit television (CCTV). Seven patients (21.2%) did not use any low vision aids during their educational years. In the 26 patients who did use aids, the educational degrees attained were distributed among all the various academic levels; three patients obtained only a high school degree, seven obtained a post-graduate degree, and thirteen were either enrolled or had completed a college degree. Similar findings were obtained in the seven patients who did not use low vision aids, where two of the patients completed only high school, three completed either a bachelors or associate degree and two patients had discontinued college.

Assessment of patients' self perception of visual function during the period of their education yielded the following results: 15 patients reported a perception of poor visual function; fair visual function was reported by 14 patients and good visual function was reported by two patients. Additionally, patients were subjectively asked to assess the perception of their quality of life: nine patients described their overall quality of life as excellent; 20 as good; one as poor; and one patient was unable to give a direct answer. The patients self reported perception of visual function during school years was significantly correlated with the self-reported perception of quality of life ($\rho = 0.53$, $p = 0.002$).

The economic status of the patients' families was found to be lower class (n=16), middle-class (n=7) and upper-class (n=9). All patients who attained only a high school degree belonged to the lower economic status group and similarly the majority of the patients that attained a degree beyond bachelors were found to be in the same lower economic group (5 out of 7). Thus, the financial status of a patient's family and the highest level of education attained were not significantly correlated ($\rho=0.11$, $p=0.56$) Among the 31 patients, 18 received financial support from sources other than their families (grants, scholarships, student loans from disability services etc.) during their educational years, while 13 did not receive any financial support apart from their families.

At the time of the interview, 20 patients were currently employed; 13 patients were full-time and seven patients held a part-time position. Ten patients were currently unemployed and one was retired. Of the 10 patients who were unemployed, six reported that it was related either partly or entirely to their visual impairment, while four reported that it was unrelated to their vision.

DISCUSSION

We performed a survey of the educational levels attained by Stargardt patients with clinically significant visual impairment during their educational years. Twenty one of 33 patients (63.6%) had achieved a degree beyond high school. This result is similar to that of a previous report where the educational levels attained by patients with Leber congenital amaurosis (LCA) were evaluated and it was found that 65% of the study population had

attained at least a college degree.⁷ Although the nature and severity of visual impairment in LCA patients differs from those with Stargardt disease, the educational levels attained were similar. LCA patients experience visual loss from birth, while those with Stargardt disease usually have a juvenile onset of visual loss. Both studies suggest that visual impairment from an early age does not preclude attainment of educational degrees beyond high school. Additionally, in patients with Stargardt disease, no significant relationship was found between the age at which patients reached a clinically significant level of low vision and the patients' level of education.

A comparison of the education levels attained by those patients who did use low vision aids (78.8%) versus those that did not (21.2%), showed no apparent trend as to the impact of the aids as they pertained to the levels of academic achievement.

A previous study from a general population of visually impaired or legally blind subjects showed an inverse association between VA loss and educational level attained.⁸ It would have been of interest to determine the relationship between our patients' visual acuity during their educational years and the highest level of education attained, however, meaningful correlations could not be obtained in the present study because of the limited distribution of VA values (i.e. only ten of the 33 patients had visual acuity better than 20/200). Furthermore, a comparison of the educational levels attained by our study population with those attained by non-visually impaired individuals over the age of 25 years (reported in 2009, U.S. Census)⁹ showed that 44% of the non-visually impaired population did not attend college, 45% had a college education, and 11% obtained a degree beyond bachelors, while in our study population, 18% did not attend a college, 61% attained a college education and 21% attained a degree beyond bachelors. Interestingly, a greater percentage of our Stargardt cohort attended a college, as compared to the general population. Nonetheless, a definitive judgment from this comparison cannot be made since our sample size was small.

Nevertheless, it is of interest that self-reported perception of visual function during school years was significantly correlated with self-reported perception for quality of life. In those patients that we were able to query regarding their quality of life (31 patients), all except two patients rated their life as either good or excellent. One patient was unable to give a direct answer, while an additional patient rated their quality of life as poor. Of interest, this patient was known to suffer from manic depression.

A substantial number of patients in our study were considered to belong to a relatively lower economic class (16/31). It is unclear whether our criteria for categorizing individuals in this economic class may have impacted upon our correlation of economic class with educational levels attained since five of the seven patients with a degree beyond bachelors were found to be in this economic group, as well as the five patients who obtained only a high-school degree. Of note, our definition of the lower economic status included family incomes of up to \$40,000 per year, which is approximately twice the poverty line for a family of four as defined by the US census bureau for 2009.⁹ Setting a lower threshold for the middle economic class may have resulted in a stronger relationship between economic class and educational level achieved. Our findings regarding economic status and educational levels attained differ from those found in a similar study of LCA patients⁶ where the authors reported that a high degree of academic achievement was attributed to the fact that the majority of their patients were considered as middle-class and therefore had better access to learning centers.

The aim of our study was to acquire insight into the education levels that patients with Stargardt disease and clinically significant visual loss can achieve. It seems that an individual's determination for attaining an education can overcome the potential impediment

imposed by impaired visual acuity. The findings reported should provide useful information for visual practitioners who counsel Stargardt patients and their parents as to the potential for accomplishing a high degree of academic achievement. Knowledge of additional factors which may contribute to academic success in these patients would benefit from further study of a larger group of patients from various economic, ethnic, religious and geographic backgrounds.

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TABLE 1

Highest Educational Level Attained by Stargardt Patients

Level of Education	No. of Patients (n = 33)	%
Not completed high school	1	3.0
High school degree only	5	15.1
Associate degree only	5	15.1
Bachelors degree		
Degree completed	8	24.2
Currently enrolled	3	9.1
Discontinued after 1 yr	1	3.0
Discontinued after 2 yrs	2	6.1
Discontinued after 3 yrs	1	3.0
Masters degree	6	18.2
Doctoral degree	1	3.0