Supplementary Online Content

Strauss RW, Kong X, Ho A, et al; ProgStar Study Group. Progression of Stargardt disease as determined by fundus autofluorescence over a 12-month period: ProgStar report No. 11. *JAMA Ophthalmol*. Published August 1, 2019. doi:10.1001/jamaophthalmol.2019.2885

This supplementary material has been provided by the authors to give readers additional information about their work.

eMethods 1. Grading of atrophic lesions on fundus autofluorescence

Grading of atrophic lesions on fundus autofluorescence

De-identified images were sent from the participating sites to a central reading center (Doheny Imaging Reading Center; DIRC) where the images were graded by two independent graders including adjudication processes by a senior investigator where applicable as previously described in detail.[1] Images with insufficient quality (ungradable images) were excluded from analysis.

Qualitative parameter

Background FAF was defined as either normal (homogeneous) or irregular (heterogeneous) as previously published: [1,2] a normal background FAF is characterized by an even, smooth distribution of FAF (homogeneous) with typical decrease at the fovea due to the presence of macular pigment, while any irregularities such as speckled, mottled or reticular patterns have been considered as irregular background FAF (heterogeneous).

Qualitative grading parameters included: the absence or presence of flecks both within and outside the arcades; presence of increased FAF at the edge of a lesion of decreased FAF (areas of increased autofluorescence were determined by comparison of the FAF of the region of interest to the background FAF); unifocal versus multifocal lesions at baseline. In addition, the presence of flecks outside the arcades was also assessed using information from clinical examination.

eMethods 2. Statistical methods

Linear models with generalized estimating equations were used to estimate the associations of baseline DAF lesion areas with participant characteristics. Longitudinally, linear mixed models (LMM), with time as the independent variable, were used to estimate the yearly change for each outcome. The models include random effects for the intercept and the slope for time which take into account the potential correlation between eyes and the correlation between repeated measurements of the same eye. The rate of lesion area change associated with each variable was first estimated in an univariate analysis. Multivariate analysis was further run by including variables that were significantly associated with the rate of area change with $p<0.1$ in univariate analyses.
Exploratory inspection of lesion growth over time showed a strong dependency on lesion size at the first visit in the retrospective cohort of ProgStar.[2] For both DDAF and DAF lesion area, stratified analysis by lesion size at the first visit was performed for eyes with small lesions (i.e., ≤ 1.90 mm^2), eyes with intermediate lesions (sizes larger than 1.9 mm^2 but ≤ 5.0 mm^2) and eyes with large lesions (>5.0 mm^2). All analyses were conducted using SAS 9.3, and p-values for two-sided tests are reported.

eReferences.

1. Strauss RW, Ho A, Munoz B, et al. The Natural History of the Progression of Atrophy Secondary to Stargardt Disease (ProgStar) Studies: Design and Baseline Characteristics: ProgStar Report No. 1. *Ophthalmology.* 2016;123(4):817-828.
2. Strauss RW, Munoz B, Ho A, et al. Progression of Stargardt Disease as Determined by Fundus Autofluorescence in the Retrospective Progression of Stargardt Disease Study (ProgStar Report No. 9). *JAMA Ophthalmol.* 2017;135(11):1232-1241.

eTable 1: Demographic characteristics at baseline visit of patients in the prospective ProgStar study

Characteristics	
Number of participants	259 (480 study eyes)
Age at first visit (mean (SD))	33.3 (± 15.1)
Age of onset of symptoms (mean(SD)*	22.3 (±12.9; range 4.0-64.0)
Age of onset ≤ 18 years	46.5%
Age of onset > 18 years	53.5%
Duration of disease until baseline visit (mean (SD))*	11.66 (±9.2; range 0-55 years)
Female	118 (54.4%)
Race	
White/Caucasian/M. Eastern	222 (85.7%)
Black/African	20 (7.7%)
Asian/Indian	10 (3.9%)
Other/Multiracial	2 (1.2%)
Unknown	4 (1.5%)
Vitamin A supplementation	14.3%
Former smoker	11.2%
Current smoker	13.5%

*missing for 18 patients

eAppendix. ProgStar Study Team

Required language to appear in the appropriate location of all publications using ProgStar (retrospective and prospective study) resources

The ProgStar study is supported by a contract from the Foundation Fighting Blindness. The ProgStar studies consist of the Chair's Office, nine clinics, two resource centers, and two affiliated centers with the following members:

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