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Longitudinal Changes of Fixation Location and Stability within 12 Months in Stargardt Disease: ProgStar Report No. 12

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Structured Abstract

Purpose: To investigate the natural history of Stargardt disease (STGD1) using fixation location and fixation stability.

Design: Multicenter, international, prospective cohort study.

Methods: Fixation testing was performed using the Nidek MP-1 microperimeter as part of the prospective, multicenter, natural history study on the Progression of Stargardt disease (ProgStar). A total of 238 patients with *ABCA4*-related STGD1 were enrolled at baseline (bilateral enrollment in 86.6 %) and underwent repeat testing at month 6 and 12.

Results: Outcome measures included the distance of the preferred retinal locus from the fovea (PRL) and the bivariate contour ellipse area (BCEA). After 12 months of follow-up, the change in the eccentricity of the PRL from the anatomical fovea was -0.0014 deg (95 % CI, - 0.27deg - 0.27 deg; p = 0.99). The deterioration in the stability of fixation as expressed by a larger BCEA encompassing 1 SD of all fixation points was 1.21 deg^2 (95 % CI, -1.23 deg^2, 3.65 deg^2; p = 0.33). Eyes with increases and decreases in PRL eccentricity and/or BCEA values were observed.

Conclusions: Our observations point to the complexity of fixation parameters. The association of increasingly eccentric and unstable fixation with longer disease duration that is typically found in cross-sectional studies may be countered within individual patients by poorly understood processes like neuronal adaptation. Nevertheless, fixation parameters may serve as useful secondary outcome parameters in selected cases and for counseling patients to explain changes to their visual functionality.