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Progression of Stargardt disease type 4 as measured by spectral-domain optical coherence tomography (SD-OCT) in the ProgStar-4 Study

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Purpose: To evaluate progression of loss in various retinal layers in patients with Stargardt disease type 4 (STGD4) over 24 months follow-up of SD-OCT findings Setting/Venue: Five centers in the U.S.A., United Kingdom and Germany.

Methods: Fifteen patients with molecularly confirmed STGD4 (PROM1 disease causing variants) were enrolled at five sites in the United States, United Kingdom and Germany. SD-OCT scans were obtained at baseline from a 20° x 20° scan area centered on the fovea and was repeated after 24 months using the built-in follow-up mode with gradable images available for 26 eyes; right eyes were chosen for analysis. The mean thicknesses (MT) of individual layers and the proportions of damage/tissue loss relative to the scanned area were calculated by a custom software within the central subfield (CS; 0.5mm radius) and the inner ring (IR; 0.5-1.5mm). The outer ring (OR; 1.5-3mm) was only analysed in eyes with adequate coverage of the posterior pole.

Results: There was a statistically significant change (all $p<0.05$) over 2 years in the segmented central subfield of the RPE in estimated trajectory of MT -3.87 (±6,1) µm, the HFC -6.22 (± 6.95) µm, the inner ring of the RPE -3.88 (±3,68) µm/, the HFC -5.49 (±6.39) µm/, the outer ring of the ONL -3.1 (±2.5) µm, the IS -1,43 (±1,26) µm, the total mean of the ONL -2.5 (±2.97) µm, the IS -1.7 (±1.45) µm, the intact area of the RPE of the inner ring -0,79 (±0,95) mm2, the PIS -0,23 (±0.36) mm2 and the outer ring of the ONL -0.12 (±0.09) mm2.

Conclusions: Significant loss could be detected in outer retinal layers by SD-OCT over a 24 months period in patients with STGD4. Loss of thickness and/or intact area of RPE, IS, OS and ONL may serve as potential endpoints for clinical trials that aim to slow down the disease progression of STGD4.