Test-Retest Reliability of Visual Function Assessments in Pseudoxanthoma Elasticum

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Purpose

Slowing of rod-mediated dark adaptation (DA) is emerging as an early marker of functional impairment in patients with Pseudoxanthoma Elasticum (PXE), offering potential as a metric to assess interventions aimed at mitigating disease progression. Despite its promise, the retest reliability of DA parameters in the PXE population remains unexplored. This study

evaluated the intersession repeatability of key visual function and DA parameters in PXE patients.

Methods

Genetically confirmed PXE patients enrolled in a prospective natural history study underwent comprehensive visual function assessments at baseline and two months later (NCT05662085). Tests included best-corrected visual acuity (BCVA), contrast sensitivity function (CSF; summarized as CSF-based acuity and area under the log CSF [AULCSF]), and DA parameters (final threshold and rod intercept time [RIT]). DA testing employed the MonCvONE device following a 59% rhodopsin bleach, with cyan and red Goldman V-sized stimuli (peak wavelengths: 500 nm and 647 nm, stimulus duration: 200 ms) presented at eccentricities of 8°, 15°, 30°, and 46° temporal to fixation (retinal coordinates). The 95% coefficient of repeatability (CoR) was calculated for each visual function biomarker to assess intersession reliability.

Results

Data were analyzed from 26 eyes of 26 patients (54% female) with a mean age of 50 ± 12 years. For visual function at fixation, the 95% CoRs were 0.193 logMAR for BCVA, 7.729 logMAR for CSF acuity, and 0.303 for AULCSF. For rod-mediated DA along the temporal horizontal meridian, the 95% CoRs for the rod intercept time (RIT) were 13.41, 12.22, 18.25, and 19.60 minutes at 8°, 15°, 30°, and 46° eccentricities, respectively. The final threshold exhibited 95% CoRs of 0.95, 0.77, 0.79, and 1.01 log units at the same eccentricities.

Conclusions

This study establishes the 95% repeatability coefficients for key visual function and DA parameters measured with the MonCvONE device in patients with PXE. These metrics provide a critical baseline for determining clinically significant changes in visual and rod function in future research and clinical trials.

Layman Abstract (optional): Provide a 50-200 word description of your work that non-scientists can understand. Describe the big picture and the implications of your findings, not the study itself and the associated details.