307 The Impact of Low Vision on Function and Everyday Activities

Tuesday, May 09, 2017 8:30 AM–10:15 AM
Room 308 Paper Session
Program #/Board # Range: 2479–2484
Organizing Section: Low Vision Group

Program Number: 2479
Presentation Time: 8:30 AM–8:45 AM
Symmetry of inherited eye disease
Todd E. Scheetz1,2, Adam P. DeLuca2, Nicole Tatro3,1, Benjamin P. Faga1,2, Douglas J. Oppedal3,4, Meagan A. Luse1,2, Richard G. Weleber1, Edwin M. Stone5,1.
Ophthalmology, University of Iowa, Iowa City, IA; 2Wynn Institute for Vision Research, University of Iowa, Iowa City, IA; 3Casey Eye Institute, Oregon Health & Science University, Portland, OR.

Purpose: To investigate whether three inherited retinal diseases are symmetrical enough for untreated fellow-eyes to be used as controls in clinical trials of gene- and cell-based treatments.

Methods: We retrospectively evaluated Goldmann visual fields from patients over the age of 10 years with molecularly-confirmed MYO7A-associated Usher syndrome (n=7), ABCA4-associated Stargardt disease (n=172), and CHM-associated choroideremia (n=23). All patients provided informed consent in accordance with the tenets of the Declaration of Helsinki. TruthMarker (iOS) was used to digitally trace the isopters from scanned versions of each visual field, generating an XML file. Fields were excluded from the study if the perimetrist marked the field as unreliable, or if incomplete data were collected. Visual field volumes were calculated from the traced isopters. Visual field volumes were calculated from the traced isopters using the field-area method of Weleber et al., (1986) multiplied by the target luminance values given of Christoforidis (2011).

The resulting paired data (OS and OD) from each visit were evaluated for concordance using Pearson’s r.

Results: Fields from both eyes were digitally traced from 396 independent patient visits. The resulting data showed extremely high concordance of the visual field volumes between right and left eyes (r^2=0.93; p=2.2x10^-30).

Conclusions: We have devised a method to capture quantitative functional measurements from historical, longitudinal visual field data. Our results demonstrate that the pattern of progression of these three diseases is highly symmetrical, supporting the use of fellow eyes as a control in gene- and cell-based treatment trials.

Correlation of visual field volumes between right and left eyes (R^2 = 0.93).

Commercial Relationships: Todd E. Scheetz, None; Adam P. DeLuca, None; Nicole Tatro, None; Benjamin P. Faga, None; Douglas J. Oppedal, None; Meagan A. Luse, None; Richard G. Weleber, Edwin M. Stone, None

Program Number: 2480
Presentation Time: 8:45 AM–9:00 AM
Characterizing the natural history of visual function in choroideremia using microperimetry and multimodal retinal imaging
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Purpose: Centripetal retinal degeneration in choroideremia (CHM) leads to early visual field restriction and late central vision loss. The latter marks an acute decline in quality of life but visual prognostication remains challenging. We investigate the natural history of visual function in CHM by correlating best-corrected visual acuity (BCVA) with microperimetry and multimodal retinal imaging.

Methods: BCVA, 10-2 microperimetry (MAIA), OCT and fundus autofluorescence (AF) were performed in both eyes of 56 CHM patients. Microperimetry was repeated in 21 eyes, enabling Bland-Altman analysis of repeatability. BCVA and macular sensitivity were correlated with age and inter-eye symmetry was evaluated. Since loss of fixation stability from foveal degeneration could affect visual testing, the distance from the fovea (on OCT) to the nearest edge of AF (representing edge of degeneration) was assessed as a potential confounder on BCVA or macular sensitivity.

Results: A Kaplan-Meier plot of the proportion of right or left eyes retaining 20/20 BCVA showed identical survival pattern (median survival 39yr). Macular sensitivity declined logarithmically with age (r=0.60, p=0.05) with a half-life of 14.72yr (95% CI 11.85 to 19.42). Zonal analysis showed faster decline nasal than temporal to the fovea. Inter-eye symmetry was more consistent for macular sensitivity (r=0.95, p<0.001) than BCVA (r=0.42, p=0.0006). The former had a coefficient of repeatability of 1.45dB (95% LOA +1.24 to -1.62). As the degeneration encroaches upon the fovea, linear reduction of...
both BCVA and macular sensitivity was seen such that near normal functions were measured when the fovea was +2500 μm away from the edge of AF whereas minimal detectable levels were reached by -800 μm.

**Conclusions:** In around half of CHM eyes, BCVA falls below 20/20 by age 39 accompanied by logarithmic decline in macular sensitivity. Both visual functions showed a high degree of inter-eye symmetry, particularly in early stages, indicating that the fellow eye can provide a suitable control for assessing interventions to one eye. A critical period of BCVA and macular sensitivity drop when the fovea is +2500 to -800 μm from the edge of degeneration corresponds to patient perception of progression from ‘split’ to ‘eccentric’ fixation. The findings will help to tailor visual prognosis and interpret outcomes of novel treatments such as gene therapy.

**Figure 1:** Composite map of MAIA microperimetry thresholds within the central 10 degrees of the macula in a cohort of 56 patients with choroideremia. Values in the key represent the mean threshold in each region.

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**Program Number:** 2481
**Presentation Time:** 9:00 AM–9:15 AM
**Effective dynamic range and retest-reliability of two-color dark-adapted fundus-controlled perimetry in patients with macular diseases**

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**Program Number:** 2482
**Presentation Time:** 9:15 AM–9:30 AM
**Two-Color (Red-Blue) Dark Adaptometry: Sensitivity, Specificity and Clinical Application**

Jeff C. Rabin, Brooke Houser, Carolyn Tailbert, Ruh Patel. Optometry, UIW Rosenberg School of Optometry, San Antonio, TX.

**Purpose:** Dark adaptometry (DA) is a sensitive test for diagnosis of retinal disease including retinitis pigmentosa (RP) and macular degeneration. Patients requiring flash electroretinograms (ERGs) often benefit from DA for proper diagnosis. Our purpose was to develop a new DA test administered during the 20 min. dark adaptation period of the standard flash ERG (www.iscerv.org) using alternating red and blue stimuli to bias the response in favor of cones (red) or rods (blue). We report sensitivity and specificity of this test.

**Methods:** An ERG Ganzfeld (Diagnosys, LLC) was used to measure DA in 21 normal subjects and 21 patients with retinal disease: RP/ Ushers syndrome, cone dystrophy, Bests, fundus albipunctatus, DUSN, macular dystrophy, Allagie syndrome. Each subject initially

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Most people with central vision loss (CVL) report difficulty watching video content experienced by people with CVL. When viewing the NV scanpath restricted-area clips, subjects with CVL (n=16), with normal vision (NV; n=60), and with NV that was blurred by defocus (n=15) watched from the gaze of subjects with normal vision (figure 1A). These abstracts are licensed under a Creative Commons Attribution-NonCommercial-No Derivatives 4.0 International License. Go to http://iovs.arvojournals.org/ to access the versions of record.
The content guide dynamically directs attention to areas that were fixated by the majority of normally-sighted viewers. A) Kernel density estimate of the gaze points for this particular frame. B) Illustration of the content guide as it appeared for the same frame.

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