Abstract: 175

Baseline Microperimetry and SD-OCT measures in the RUSH2A study

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Purpose:
To address limited data characterizing retinal degeneration related to disease-causing sequence variants in Usher syndrome 2A (USH2A), the Foundation Fighting Blindness Consortium is conducting a 4-year multicenter, international natural history study titled Rate of Progression in USH2A-related Retinal Degeneration (RUSH2A). We report baseline data on mesopic microperimetry (MP) and spectral domain optical coherence tomography (OCT) and their relationships with other baseline characteristics.

Methods:
General linear models were used to assess the association between demographic and clinical characteristics, including gender, race, enrollment age, disease duration, smoking, use of dietary supplements, MP mean sensitivity and OCT ellipsoid zone (EZ) area. The association between MP mean sensitivity and OCT EZ with other measures (including visual acuity (VA), and central subfield thickness (CST) within the center 1mm) was assessed using Spearman correlation coefficients.

Results:
All participants (N=127) had OCT measures, while MP was obtained at selected sites (N= 93). Comparing participants with Usher syndrome type 2 (USH2, N=80) to autosomal recessive non-syndromic RP (ARRP, N=47), the central structural measures of OCT EZ (3.1±5.7 mm² vs 4.3±5.6 mm², p=0.26) and CST (253.1±57.5 µm vs 263.6 ± 32.9 µm, p=0.26) were similar. Mean MP sensitivity was similar by diagnosis (5.4 ± 4.9 dB vs 6.7± 5.1 dB, p=0.22). Longer disease duration was associated with smaller OCT EZ (p<0.001) and lower mean sensitivity (p=0.01), adjusted for clinical diagnosis. Better VA, larger OCT EZ area, larger CST were associated with greater MP mean sensitivity (r>0.3 and p<0.01). Better VA and larger CST were associated with larger OCT EZ area (r>0.6 and p<0.001).

Conclusions:
The baseline RUSH2A data revealed similar structural and functional metrics between the USH2 and ARRP participants. Longer disease duration was associated with more severe abnormalities of retinal structure and function, adjusted for clinical diagnosis. MP and OCT measures may provide useful metrics to monitor during disease progression in studies of USH2A-related retinal degeneration.