A cross-sectional and longitudinal study of retinal sensitivity in RPE65-associated Leber congenital amaurosis – Supplementary Material

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Supplementary Figure Legends

Supplementary Figure S1: Comparison of different visualizations and indices of static perimetry exam from MM_0262. (A) A plot demonstrating the 164-point grid used in this study. (B) Incremental color scale plot generated by the Octopus 900 (Haag-Streit, Köniz, Switzerland). (C) Superior oblique view of the 3-dimensional hill of vision generated by
VFMA, with iso-sensitivity contours every 2dB intervals. (D) En face view with selection boundaries for total volume ($V_{TOT}$), central 30° volume ($V_{30}$) and central 15° volume ($V_{15}$) shown.

Supplementary Figure S2: Total visual field sensitivity, $V_{TOT}$, compared to mean sensitivity. Shown is a scatter plot of $V_{TOT}$ of the left eyes ($n = 18$) of all patients against mean sensitivity, with the corresponding linear regression line. This demonstrates a strong, positive linear correlation ($r = +0.98$) between total visual field sensitivity and mean sensitivity.
Supplementary Figure S3: Total visual field sensitivity, $V_{TOT}$, compared to age. Shown is a scatter plot of $V_{TOT}$ of the left eyes (n=18) of all patients against age, with the corresponding linear regression line. This demonstrates a moderate, negative linear correlation ($r = -0.58$) between total visual field sensitivity and age.
Supplementary Figure S4: Central 15° visual field sensitivity, $V_{15}$, compared to age. Shown is a scatter plot of $V_{15}$ of the left eyes (n=18) of all patients against their age, with the corresponding linear regression line. This demonstrates a moderate, negative linear correlation ($r = -0.55$) between central 15° visual field sensitivity and age.
Supplementary Figure S5: Central 30° visual field sensitivity, $V_{30}$, compared to best corrected visual acuity. Shown is a scatter plot of $V_{30}$ of the left eyes ($n=18$) of all patients against best corrected visual acuity, with the corresponding linear regression line. This demonstrates a weak linear correlation ($r = -0.27$) between central 30° visual field sensitivity and best corrected visual acuity.
Supplementary Figure S6: Central 30° visual field sensitivity, $V_{30}$, compared to contrast sensitivity. Shown is a scatter plot of $V_{30}$ of the left eyes (n=18) of all patients against contrast sensitivity, with the corresponding linear regression line. This demonstrates a moderate linear correlation ($r = +0.46$) between central 30° visual field sensitivity and contrast sensitivity.
Supplementary Figure S7: Change in total visual field sensitivity, $V_{TOT}$, over time in a subset of 13 patients. Shown is a line graph of serial $V_{TOT}$ measurements in subjects over time. Subjects have also been color-coded by genotype group (red: mild, blue: intermediate and green: severe). This demonstrates the slow loss of total retinal sensitivity seen in this cohort of RPE65-LCA.
Supplementary Figure S8: Change in central 15° visual field sensitivity, \( V_{15} \), over time in a subset of 13 patients. Subjects have also been color-coded by genotype group (red: mild, blue: intermediate and green: severe). Shown is a line graph of central \( V_{15} \) measurements in subjects over time. This demonstrates the slow loss of the central 15° of retinal sensitivity seen in this cohort of patients with \textit{RPE65}-LCA.

**Supplementary Tables**

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<thead>
<tr>
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<th>Coefficient of repeatability</th>
<th>Confidence interval</th>
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<tbody>
<tr>
<td>( V_{\text{Tot}} )</td>
<td>20.98</td>
<td>±0.40</td>
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<tr>
<td>( V_{30} )</td>
<td>3.61</td>
<td>±0.07</td>
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<tr>
<td>( V_{15} )</td>
<td>0.91</td>
<td>±0.02</td>
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Supplementary Table S1: Table showing the coefficient of repeatability and its confidence interval for total visual field sensitivity (\( V_{\text{Tot}} \)), Central 30° visual field sensitivity (\( V_{30} \)), and central 15° visual field sensitivity (\( V_{15} \)).
Discussion of Supplementary Materials

The above supplementary materials support our finding that retinal sensitivity using static perimetry quantified with VFMA software is an important measure of visual function.¹ We demonstrate the radially-designed, centrally condensed grid with the 164 test locations used, the Octopus 900 (Haag Streit, Köniz, Switzerland) print out, and how this is modelled into a hill of vision and the subsequent volumetric analysis. The figures in this section further demonstrate the correlation of the volumetric metrics with other indices of retinal function. Furthermore, Supplementary Table S1 shows the coefficient of repeatability for the cohort of subjects tested.

References