Progression of Vision Loss in Macular Telangiectasia Type 2

Tjebo F.C. Heeren,¹ Traci Clemons,² Hendrik P.N. Scholl,³ Alan C. Bird,⁴
Frank G. Holz,¹ Peter Charbel Issa¹*

¹ Department of Ophthalmology, University of Bonn, Germany
² The EMMES Corporation, Rockville, Maryland, United States
³ Wilmer Eye Institute, Johns Hopkins University School of Medicine, Baltimore, Maryland, United States
⁴ Moorfields Eye Hospital NHS Foundation Trust and UCL Institute of Ophthalmology, London, United Kingdom

Supplementary Material
Supplementary Methods

Modified version of the classification proposed by Gass and Blodi¹:

Stage 0  No obvious disease manifestations, but fellow eye with stage 1-5
Stage 1  Diffuse hyperfluorescence in late phase fluorescein angiography
Stage 2  Reduced parafoveolar retinal transparency
Stage 3  Dilated right angled venules
Stage 4  Intraretinal pigment clumping
Stage 5  Neovascular complex

¹ Idiopathic juxtafoveolar retinal telangiectasis. Update of classification and follow-up study. Ophthalmology 1993;100(10):1536-46

Supplementary Results

Baseline characteristics of the study cohort

Forty patients (22 female, 18 male; all Caucasians) with MacTel type 2 were included in the study. Nine eyes were excluded, resulting in 71 eyes remaining for analysis (see flow chart below). Three eyes that underwent cataract surgery during the observation period showed no obvious deviation from the disease course in other eyes and were not excluded from analysis.

Flow chart of exclusion and inclusion of all analyzed eyes.
Supplementary Table S1: Details of previous therapies in the study cohort.

<table>
<thead>
<tr>
<th>Previous treatment of included eyes</th>
<th>n (eyes)</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>48</td>
</tr>
<tr>
<td>Focal Argonlaser-Coagulation (ALC)</td>
<td>6</td>
</tr>
<tr>
<td>Photodynamic Therapy (PDT)</td>
<td>1</td>
</tr>
<tr>
<td>Intravitreal Ranibizumab</td>
<td>5</td>
</tr>
<tr>
<td>Intravitreal Bevacizumab (IVB)</td>
<td>4</td>
</tr>
<tr>
<td>Intravitreal Triamcinolon (IVT)</td>
<td>3</td>
</tr>
<tr>
<td>IVB+IVT+ALC</td>
<td>2</td>
</tr>
<tr>
<td>IVT+PDT</td>
<td>1</td>
</tr>
<tr>
<td>IVT+ALC</td>
<td>1</td>
</tr>
</tbody>
</table>

61% of treated eyes had a scotoma at baseline compared to 40% of untreated eyes. 83% of treated eyes revealed progression of a scotoma compared with 46% of untreated eyes. Conclusions from these ratios need to be interpreted with caution because of possible confounding factors. For instance, it may be that the worse eye was treated more frequently than the better eye, potentially resulting in higher frequency of scotomata in treated eyes.

Supplementary Figure S1: Analysis of microperimetric mean sensitivity

*Left:* Analysis of the mean sensitivity (±SD) within the central testing field (central 4°x8°) revealed a minor but significant decrease during the review period in eyes with new development or progression of a scotoma (13.9dB±3.6 to 12.4dB±3.2; \( P=0.0001 \)) and showed no change in those without scotoma progression (18dB±1.3 to 17.9dB±1.4; \( P=0.67 \)).

*Right:* Both groups showed no change of mean sensitivity outside the central testing field (17.2dB±1.5 to 17.1dB±1.9 and 17.8dB±1.2 to 17.7dB±1.5, respectively; \( P=0.43 \) and 0.77). At baseline, eyes with and without growth of an absolute scotoma differed significantly in mean sensitivity of the central testing field (\( P<0.0001 \)), but not of the outer testing field (\( P=0.10 \)).
Supplementary Figure S2: Functional change on microperimetry and visual acuity testing based on visual acuity level at baseline

Baseline best corrected visual acuity (BCVA) level (≤20/50 versus >20/50) was not predictive for the relative frequency of functional decline on longitudinal microperimetry testing (new/additional test point showing an absolute scotoma) or of a significant decrease in BCVA (>2 lines).
Supplementary Figure S3: Microperimetry examinations and associated clinical findings of two eyes that developed an absolute scotoma

A: The small lesion in the photoreceptor layer visible on optical coherence tomography (OCT) at baseline was not associated with a detectable scotoma on microperimetry testing. No marked changes are visible on fundus photography and late phase fluorescein angiography in the follow up compared to baseline. However, OCT revealed an increased lesion size at the level of the photoreceptor layer as structural correlate of the incident scotoma.

B: Representative example of changes on OCT imaging observed in eyes with the development or increase of an absolute scotoma. The loss of tissue in the photoreceptor layer in the follow-up examination is associated with an absolute scotoma on microperimetry testing.
Supplementary Figure S4: Change on visual acuity and microperimetry testing in individual eyes.

The black columns represent the number of testing points with a new/additional absolute scotoma (scale=right Y-axis) on microperimetry testing. The grey columns represent change of best corrected visual acuity (BCVA) (scale=left Y-axis). The shaded area marks the range of stable BCVA (±2 lines). There are 5 eyes that decreased in BCVA in absence of changes on microperimetry testing. Seven eyes showed a decrease in BCVA and a new development or progression of an absolute scotoma. The 2 eyes with the worst progression on microperimetry testing show stable BCVA, as the scotoma already affected the foveal center and the BCVA was already low.