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Released: 07/15/2025
Valid until: 07/15/2026
Time needed to complete: 1h 03m

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Anatomy Meets Function – Correlating Imaging with Symptoms

Dr. Wykoff:
Hi, this is CME on ReachMD. I'm Charles Wyckoff, and it's a pleasure to be here with good friend and colleague, Dr. Dean Elliott.

So, Dean, my question for you is, we know that in patients with macular telangiectasia type 2, or Mac-Tel, visual acuity does not often correlate well at a patient level with visual function.Are there anatomic findings that we should be aware of that better correlate with vision loss in Mac-Tel?

Dr. Elliott:
Thanks for having me, Charlie. That is a great question.Distance vision, as measured by BCVA, tends to be maintained in patients with Mac-Tel.Gass and Blodi found in a review of 140 cases spanning 28 years, that only 33% of patients had visual decline to less than 20/200. Another study showed 25% of patients maintained stable vision over 17 years. And BCVA only decreases once photoreceptor and ellipsoid zone loss occurs at the fovea.

Functional vision loss is more common than decreases in BCVA. Patients often have paracentral visual defects, as shown in the pie chart. Reading speed is often reduced. Reading ability requires at least 4 degrees of preserved central vision to process long sentences, and reading speed can be used to assess visual function.

Microperimetry is also used in clinical trials. It's good at assessing paracentral vision. Scotomas are sharply demarcated and profound, and the presence of scotomas at baseline are predictive of functional vision loss, as shown in the graph.

Microperimetry defects correlate significantly with photoreceptor loss in SDOCT and en face OCT, and these OCT modalities are great tools for measuring ellipsoid zone loss. Can you tell us why, Charlie?

Dr. Wykoff:
Here's a great example. It illustrates exactly what you were just describing, Dean. On the left is an image from 2020, over to 2024 on the right. And if you look at the cross-sectional B scan at the bottom, or the en face, ellipsoid zone, loss zone on the top, those black areas where the EZ is being lost. And critically, visual acuity here remained relatively stable; 20/30 on the left, 20/30 on the middle, but then 20/40 on the right. All in the same ballpark. And yet, if you look at the EZ zone loss, there's significant increase overtime correlating with decreased functional vision in many cases, even though the visual acuity itself is relatively stable.

So, overall, we know that the probability of non-central ellipsoid zone loss due to Mac-Tel is about 76% among these patients, and patients with non-central EZ loss have a 45% chance of central EZ loss over time. In comparison, loss of BCVA occurs in only about 15 to 25% of patients, so ellipsoid zone loss, again, much more common, progressive ellipsoid zone loss is more common also, than visual acuity. Significant loss. Progression of the area of ellipsoid zone loss en face OCT is the best imaging endpoint for Mac-Tel. We believe, currently, en face OCT analyses have been accepted, of course, by the FDA as primary endpoint for Mac-Tel as demonstrated with the

recent FDA approval.

We can also see retinal cavitations in patients Mac-Tel, and my question for you, Dean, is how do these features relate to visual decline?

Dr. Elliott:
Cavitations are a feature of Mac-Tel. They may be mistaken for macular hole. They tend to fluctuate with time. These are not pathognomonic for Mac-Tel, but they're very suggestive. Volume of these cavities negatively correlates with BCVA, but not the area of ellipsoid zone loss.

Dr. Wykoff:
Overall, in summary, both spectral domain and en face OCT are great for imaging Mac-Tel. They can track disease progression. Ellipsoid zone loss correlates with visual function loss more than pure best corrected visual acuity. Functional vision loss is what patients with Mac-Tel struggle with the most, in discussions with them.And we'll talk more about functional vision loss and the impact on quality of life in our next episode.

For now, that is all the time we have time for. Thank you, Dean, for your participation here.

Dr. Elliott:
Thank you.