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### Reply

#### Dear Editor:

We thank Dr. Oguz for his interest in our review article, which was intended to cover current aspects of endophthalmitis management. The scope of our article required that we limit our discussion to antimicrobial agents of immediate applicability to this condition. Nevertheless, we strongly agree that agents such as taurolidine may have great promise and deserve further investigation as to their value in the clinical management of endophthalmitis.

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#### Dear Editor:

Idiopathic juxtafoveal retinal telangiectasis is a clinical entity distinctly different from secondary telangiectasis, which can result from various diseases. Group 2a acquired idiopathic juxtafoveal retinal telangiectasis (IJRT) is the most common group and has clinical, optical coherence tomographic (OCT),<sup>1,2</sup> and fluorescein angiographic (FA)<sup>3</sup> findings unlike other macular diseases. The pathophysiology of group 2a is controversial. Gass initially suggested a primary role of the leaky retinal capillaries with subsequent chronic nutritional damage to Muller cells,<sup>3</sup> and later, commented that “this disorder is not primarily a leaky retinal blood vessel disease,” but rather “the primary abnormality may reside in one or both of the parafoveal retinal neural or Muller cells.”<sup>4</sup> FA in these eyes usually reveals temporal parafoveal telangiectatic vessels and intraretinal fluorescein leakage that spares the foveal center.<sup>3</sup> Cohen et al<sup>1</sup> recently reported on the OCT findings in a large population of group 2a idiopathic juxtafoveal retinal telangiectasis, and they proposed that Muller cell dysfunction may explain the clinical, FA, and OCT findings in these patients. Given that Muller cells confer barrier properties to the retinal capillary endothelium and help regulate

retinal blood flow,<sup>5,6</sup> they believe that primary degeneration or dysfunction of Muller cells would be accompanied by a breakdown of the blood–retinal barrier (alterations in the parafoveal retinal capillaries) in group 2a idiopathic juxtafoveal retinal telangiectasis.<sup>5</sup> Moreover, they believe that the outer retinal atrophy seen with OCT could not be caused by retinal vascular abnormalities alone, because the outer retina derives oxygen and nutrients from the choriocapillaris, and that, again, Muller cell dysfunction could be responsible for outer retinal atrophy and degeneration, because Muller cells maintain the health of the surrounding neurons including the outer retinal neurons. Interestingly, given that the intraretinal voids seen in group 2a idiopathic juxtafoveal retinal telangiectasis are unlike those seen in patients with macular edema caused by retinal vein occlusion, diabetes, and inflammation, because not associated with increased retinal thickness,<sup>2</sup> they proposed that the characteristic localized patches of fluid seen in group 2a patients may be caused by fluid that leaks from the parafoveal retinal capillaries and migrates to the fovea, and that such proteinaceous fluid could accumulate within the foveal avascular zone because of the lack of a capillary system there to remove them.

Recently, Moon et al<sup>7</sup> have shown favorable short-term results using bevacizumab in the treatment of a patient with group 2a idiopathic juxtafoveal retinal telangiectasis. Of note, the authors, in their report, did not show FA, and even OCT scans reported are quite different from the typical scans of group 2a idiopathic juxtafoveal retinal telangiectasis patients (occult foveal cysts without increased foveal thickness).<sup>1,2</sup> Moreover, the authors did not exclude the presence of diabetes and prediabetes, which could represent an occasional cause of group 2a idiopathic juxtafoveal retinal telangiectasis.

We also decided to assess the effects of intravitreal bevacizumab on a patient with nonproliferative group 2a idiopathic juxtafoveal retinal telangiectasis, and we performed the same treatment as Dr Moon. Indeed, in our patient, intravitreal bevacizumab injection failed to lead to anatomic or visual amelioration.

Therefore, we agree with Cohen et al<sup>1</sup> and we believe that bevacizumab injection would be ineffective in group 2a idiopathic juxtafoveal retinal telangiectasis, because of the primary degeneration or dysfunction of Muller cells in such patients, which would be not VEGF-related.

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### Reply

#### Dear Editor:

By revealing subtle structural retinal abnormalities, optical coherence tomography (OCT) has increased our understanding of many macular disorders. Several recent publications have described the unique OCT findings in eyes with group 2a idiopathic juxtafoveal retinal telangiectasis (IJRT).<sup>1–6</sup> In our recent study published in *RETINA*, we suggested that the unusual OCT abnormalities and the other clinical features of group 2a IJRT might be attributable to Muller cell dysfunction.<sup>1</sup> My coauthors and I appreciate the letter by Drs. Querques and Delle Noci in support of our premise that Muller cell dysfunction may be the primary cause of group 2a IJRT. They base their support of the Muller cell dysfunction hypothesis on their group 2a IJRT patient's failure to respond to intravitreal bevacizumab injection.

The authors raise several key issues regarding the pathophysiology, diagnosis, and treatment of nonproliferative group 2a IJRT. Below I discuss where I agree and disagree with Drs. Querques and Delle Noci on these three important points.

When treating any patient, an understanding of the pathophysiology of their disease is helpful. Nevertheless, we can successfully treat many patients with poorly understood disorders. For example, there are effective treatments for patients with Eales' disease and idiopathic retinal vasculitis, aneurysms, and neuroretinitis even though the etiology and pathophysiology of these disorders remains unknown. Patients with these diseases develop nonperfusion and proliferative

retinopathy. Because we know that panretinal photocoagulation prevents vision loss in eyes with nonperfusion and proliferative retinopathy, we can prevent vision loss in patients with these mysterious diseases with panretinal photocoagulation.<sup>7,8</sup> Conversely, some diseases we understand pretty well respond to therapies that seem like they should not work. For example, macular edema from retinal capillary dysfunction in patients with retinal vein occlusion and diabetic retinopathy has traditionally been attributed to structural capillary defects caused by these diseases. Yet recent studies have shown that macular edema caused by these disorders responds favorably to VEGF inhibition by intravitreal bevacizumab suggesting that there may be a biochemical cause of the macular edema.<sup>9,10</sup>

While I appreciate Drs. Querques and Delle Noci agreeing with my coauthors and me that Muller cell dysfunction might be the primary cause of group 2a IJRT, I do not agree that Muller cell involvement precludes potential usefulness of bevacizumab in these patients. Patients with group 2a IJRT develop leaky parafoveal capillaries that may contribute to vision loss in these eyes. Muller cells could cause retinal capillaries to leak because they synthesize and secrete vascular endothelial growth factor (VEGF).<sup>11</sup> Since it is possible that leaky parafoveal capillaries contribute to vision loss, and it is possible that the parafoveal capillaries leak because of excess VEGF, it is also possible that VEGF inhibitors like bevacizumab could help patients with group 2a IJRT. A study measuring intraocular VEGF levels in eyes with group 2a IJRT could determine whether VEGF plays a role in this disease.

In addition to its pathophysiology, Drs. Querques and Delle Noci discuss the important problem of accurately determining if a patient has group 2a IJRT. I agree with the authors that this disease is difficult to diagnose. Group 2a IJRT is rare. It usually presents with subtle signs and symptoms. During my medical retina fellowship at the Bascom Palmer Eye Institute in 1992–1993, I had the opportunity to review fluorescein angiograms with Dr. Gass that were sent to him by retinal specialists from around the world as “unknowns.” Many of these mail-in consults were of eyes with group 2a IJRT.

In my community, I have seen patients with group 2a IJRT incorrectly diagnosed and treated by retina specialists for dry age-related macular degeneration, pseudophakic cystoid macular edema, diabetic macular edema, and macular pucker. Although eye vitamins for macular degeneration and topical anti-inflammatory for pseudophakic cystoid macular edema have minimal risks, steroid injections, focal laser, and vitrectomy surgery have significant risks. One eye with

group 2a IJRT that was misdiagnosed with macular pucker had vitrectomy surgery with epiretinal membrane peeling and developed vision loss from a postoperative macular hole. Another eye with group 2a IJRT and vision of 20/50 that had grid laser, intravitreal Kenalog, and vitrectomy surgery with epiretinal membrane peeling developed permanent vision loss to hand motion vision from an episode of postvitrectomy elevated intraocular pressure. Although there is no current effective treatment for eyes with nonproliferative group 2a IJRT, at least with a proper diagnosis, these patients can be spared potentially harmful interventions.

Finally, since Drs. Querques and Delle Noci are concerned with the ineffectiveness of bevacizumab injection in their patient, it is worthwhile outlining several problems to consider when assessing potential treatments for patients with group 2a IJRT. First, most patients with this disease maintain good vision in at least one eye.<sup>12</sup> Of the 22 patients we reported in our study, 16 (73%) had 20/40 vision or better in at least one eye.<sup>1</sup> The benign natural history of this disease in the majority of patients should be carefully weighed against potential risks of any proposed therapy. Second, when vision loss occurs in eyes with nonproliferative group 2a IJRT, it does so gradually, usually over several years. Since vision loss is uncommon and gradual, a commitment to long-term treatment would be needed to show a lasting benefit. Third, we have learned from OCT that vision loss in eyes with group 2a IJRT is often associated with outer retinal atrophy. There is no treatment for retinal atrophy. If, in some cases, retinal atrophy is secondary to chronic subretinal fluid, like in patients with chronic central serous chorioretinopathy, then eliminating subretinal fluid in these patients before the retina has been irreversibly damaged might be helpful. The elimination of intraretinal fluid may or may not also be beneficial. In our study OCT revealed intraretinal or subretinal cysts in 23 of 41 eyes.<sup>1</sup> Because of the above-mentioned considerations, there would be many obstacles to overcome in designing a treatment trial for this mostly benign, relatively uncommon retinal disease that sometimes causes gradual vision loss.

We thank the authors for their interest in our article. Although group 2a IJRT is a rare disorder, its unique constellation of clinical and OCT findings makes it worthy of further study. Perhaps a better understanding of this enigmatic disease will reveal secrets about the retina that could benefit many.

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