

ABOUT THE AUTHORS



MARK GREVE, MD: Dr. Greve attended and received his Doctor of Medicine in 1983-1988 from the University of Saskatchewan. He then began his Ophthalmology Residency in 1988, at the University of Saskatchewan, completing his Residency in 1992. Dr. Greve spent the next two years, till 1994, completing his Medical/Surgical Retina Fellowship at Louisiana State University. Dr Greve is currently an Associate Clinical Professor with the University of Alberta and is Chief of Vitreoretinal Surgery.



MARK SEAMONE, MD: Dr. Seamone completed a 4-year BSc in Biology at Acadia University. Following his undergraduate, he completed a MSc in Immunology and his Doctor of Medicine as a member of the Leaders in Medicine Program, University of Calgary. He then completed a 5-year Ophthalmology residency at Dalhousie University. Dr. Seamone concluded his training by completing a 2-year medical and surgical Vitreoretinal fellowship at the University of Alberta. He is a Fellow of the Royal College of Physicians and Surgeons of Canada. He is an Assistant Clinical Professor in the Department of Ophthalmology, University of Alberta, and serves as the Fellowship Director for the University of Alberta Vitreoretinal Fellowship Program.



CARL SHEN, MD: Dr. Shen completed his Bachelor of Medical Sciences at Western University followed by his Doctorate of Medicine from the University of Toronto and subsequent residency training in ophthalmology at McMaster University. He is currently completing his vitreo-retinal fellowship at the University of Alberta.

The evolving role of OCT in pathologic myopia

Carl Shen, MD Mark Seamone, MD Mark Greve, MD

Introduction

The global burden of myopia represents a significant public health concern that is expected to continue to increase in the near future. It is estimated that 50% of the world's population will be affected by myopia by 2050¹, with a disproportionately high prevalence in Asia. High myopia, where the spherical equivalent refractive error is equal to or higher than 6.00 diopters, is expected to increase in prevalence from 2.7% to 10% during this period¹. The severity of myopia is of paramount concern to clinicians as higher levels are associated with pathologic myopia (PM) and increased risk of vision loss. Pathologic myopia, as recently defined by the International Myopia Institute, is an excessive axial elongation associated with myopia that leads to structural changes in the posterior segment of the eye that can lead to loss of best-corrected visual acuity². These structural changes and their complications include posterior staphyloma, myopic choroidal neovascularization, myopic maculopathy, myopic traction maculopathy, dome shaped maculopathy, optic disc changes and glaucoma associated with myopia, and retinal detachments.

The advent of optical coherence tomography (OCT) has facilitated the characterization, diagnosis, and management of several of these complications associated with PM and will be the focus of this article. Imaging the highly myopic eye represents a crucial step in the identification of these complications and poses its own unique challenges. Researchers have demonstrated the advantage of 3D cube scans in the detection of pathology compared to 1- and 5-line rasters³. Using vertical scanning patterns aligning where the radius of curvature is larger relative to the horizontal plane of the myopic eye can minimize associated artifacts. Wide scans, facilitated by emerging technologies such as swept- source OCT and ultra wide-field OCT, are useful in cases of PM where the pathology can initiate peripherally⁴.

Myopic Choroidal Neovascularization

It is estimated that 5-11% of patients with PM will develop myopic choroidal neovascularization (MCNV)⁵. Effective treatment, primarily with anti-VEGF agents, can alter the natural history of MCNV that would otherwise result in significant vision loss.

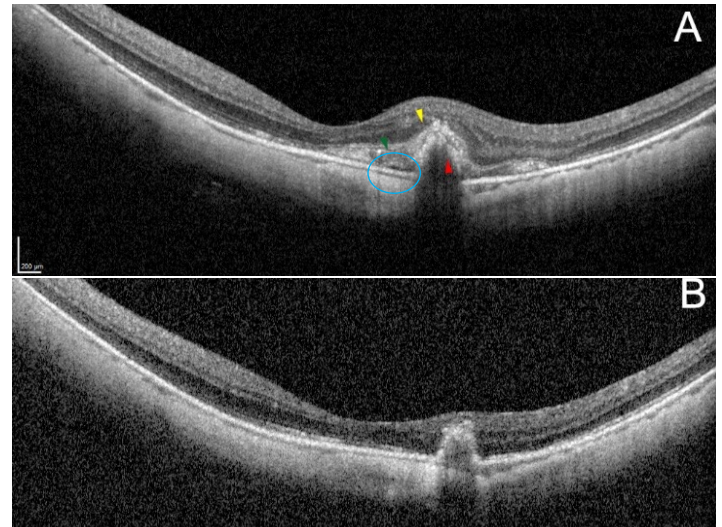


Figure 1. A) Pigment epithelial detachment (red) with overlying disruption of the ellipsoid and external limiting membrane (yellow arrow), subretinal hyper-reflective material (green arrow), and small amount of subretinal fluid (blue circle). B) Follow up OCT after 1 year of “treat and extend” anti-VEGF. Note the consolidation of the borders of the PED, and absence of subretinal fluid and sub-retinal hyperreflective material.

Diagnosis: MCNV often appears as a type 2 CNV on OCT with a hyperreflective mound above the retinal pigment epithelium (RPE) (**Figure 1**). Additional OCT features of MCNV include a loss of the ellipsoid layer and Bruch's membranes, absence of the external limiting membrane, and retinal thickening⁶. The degree of intra- and subretinal fluid by OCT is less in MCNV compared to other etiologies and thus may be a less sensitive marker for its detection if used in isolation⁷. At the same time, features such as the absence of subretinal fluid, may aid in the differentiation of MCNV from neovascularization caused by age-related macular degeneration, and inflammatory causes⁸. The agreement between OCT and reference standard fluorescein angiography (FA) in the detection of MCNV has been reported to be about 70-90%^{9,10}. Optical coherence tomography-angiography (OCT-A) may be more sensitive, being able to detect over 90% of MCNVs¹¹, and may also be useful in differentiating MCNV from hemorrhage related to lacquer cracks¹². However, to date, FA remains the gold standard for diagnosis of MCNV.

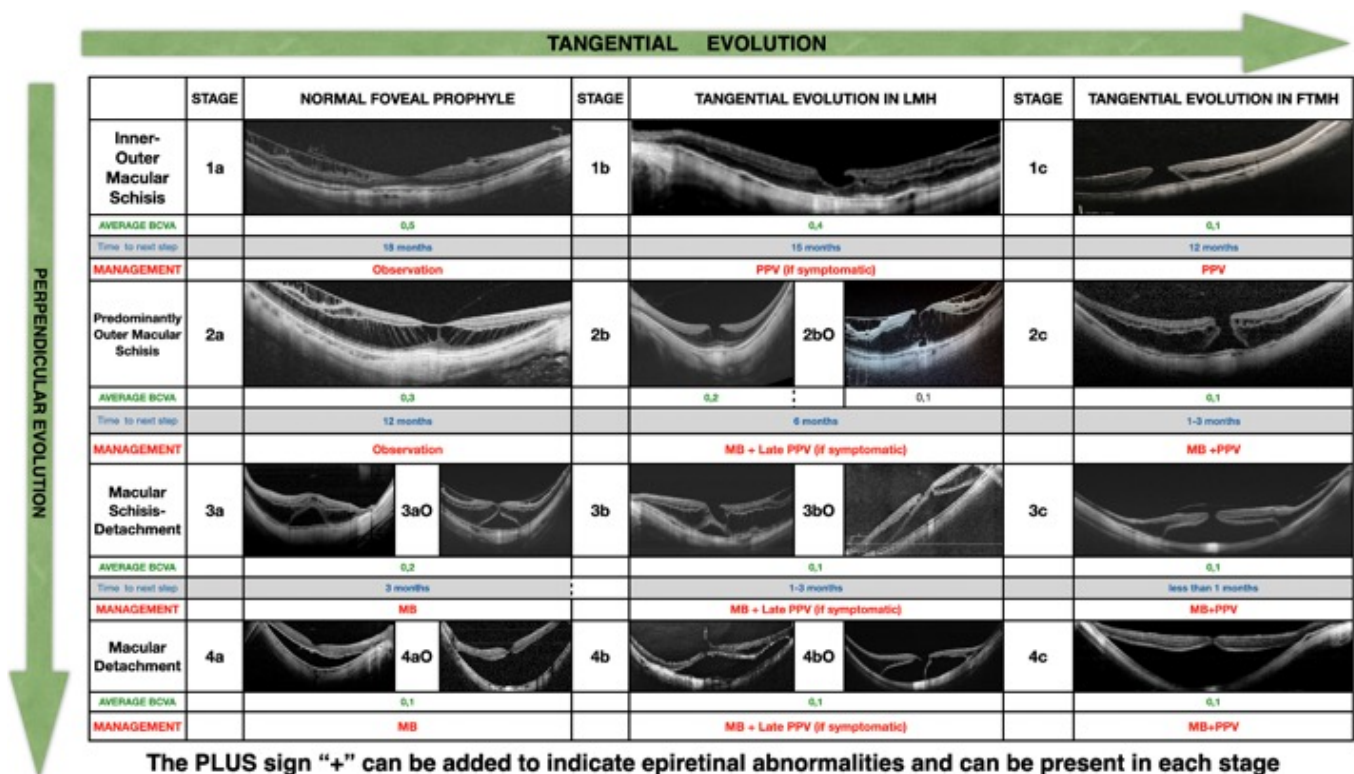


Figure 2. Myopic Traction Maculopathy Staging System based on tangential and perpendicular evolution of changes in MTM; Adpated from Parolini, B et al., 2021"

Disease Monitoring: OCT provides a rapid, non-invasive means of monitoring disease activity and response to treatment. A characteristic "fuzzy area" of the hyperreflective lesion often corresponds with active FA leakage¹³. Subretinal hyperreflective exudation is similarly predictive for MCNV activity¹⁴. Several studies have sought to identify OCT characteristics capable of prognosticating development of MCNV, response to treatment, and recurrence. Eyes with MCNV have been found to have reduced choroidal thickness compared to fellow eyes¹⁵. Thinner choroids have also been associated with recurrence of MCNV at the 1-year follow up and with a lower resolution rate after single injections of anti-VEGF therapy¹⁶.

Myopic Traction Maculopathy

Myopic traction maculopathy (MTM) refers to a collection of conditions including vitreomacular traction, epiretinal membrane, lamellar hole, macular hole, myopic foveoschisis/macular schisis that are unified by their underlying etiology of tractional forces acting on the retina in highly myopic eyes². MTM may affect up to 30% of patients with PM¹⁷. OCT findings in MTM have recently been organized into a staging system¹⁸ called the MTM Staging System (MSS) (**Figure 2**). The MSS organizes the evolution of changes from forces perpendicular to the retina (stage 1-4) with forces tangential to the retina (a-c), to arrive at a number and letter staging (e.g 2B). Stage 1

represents inner-outer macular schisis, stage 2 represents predominantly outer macular schisis, stage 3 represents macular schisis detachment, and stage 4 represents macular detachment. Stage a represents a normal foveal profile, stage b represents lamellar hole changes, and stage c represents macular hole changes. Epiretinal abnormalities are designated with a "+" and can occur at any stage.

Myopic Macular Schisis: Myopic Macular Schisis (MMS), or foveoschisis, is evident on OCT as retinoschisis in multiple retinal layers, bridged vertically by presumed Müller cells. Separation of the internal limiting membrane (ILM) from the remaining retinal layers can occur. In early stages (stage 1-2 MSS), the ellipsoid zone is usually well-preserved and visual function is minimally affected. A large natural history study of MTM¹⁹ found that over 36.2 ± 6.2 months of follow up, 11.6% (n= 24/207) of eyes experienced progression of MTM (**Figure 3**), with higher rates of progression (42.9%) in eyes with more extensive MMS.

Myopic Macular Hole: Macular holes are reported to occur in approximately 8.4% of eyes with PM²⁰. Data has shown that there is a differentiation between two types of macular holes in myopic eyes. The first is a "flat type", characterized by cystic changes at the edge of the hole and similar success rates of surgical closure similar to macular holes in non-myopic eyes²¹. The second is a "schisis type"

characterized by accompanying retinoschisis changes which are evident at the borders of the hole. These latter holes have a lower success rate of closure and are at high risk of progression to macular hole retinal detachments.

Treatment: Intervention at the MMS stage is generally guided by progressively worsening visual symptoms, metamorphopsia, and reduction in visual acuity. Treatment consists largely of surgical intervention across the spectrum of MTM and comprises vitrectomy and/or macular buckling. In a prospective, non-randomized study of 62 consecutive eyes, rates of visual improvement with surgery were shown to be greater when foveal detachment or disruption was evident²². Several variations of vitrectomy have been proposed, vitrectomy alone, with membrane peeling, with ILM peeling, ILM flaps, subretinal expansion, retinal incisions, and grafting procedures of amniotic membrane or retinal tissue. There has been renewed interest in macular buckling for the management of MTM with the hypothesis that anatomical and functional outcomes may be superior to vitrectomy alone^{23,24} (**Figure 4**). Several modifications to the standard macular buckling technique have been proposed to facilitate the surgery including modified buckle shapes²⁴ and internal chandelier placement²⁵. Intraoperative OCT has been used to augment surgery in MTM eyes through the detection of residual epiretinal membrane (ERM), ILM, and cortical vitreous and has revealed undetected holes post peeling²⁶. Clinicians have proposed a treatment algorithm based on their MSS using permutations of vitrectomy and macular buckling depending on the stage of disease, with vitrectomy better suited to address tangential traction and macular buckling better suited to address perpendicular traction²⁷. Such studies underscore the utility of OCT in cases of MTM in potentially identifying optimal treatment strategies corresponding to pathophysiologic principles.

Myopic Maculopathy

Myopic maculopathy (MM), or myopic macular degeneration, are terms often used interchangeably to describe several of the degenerative features in PM that contribute loss of best corrected visual acuity in myopia. The most widely used classification system is the meta-analysis of pathologic myopia (META-PM)²⁸ which divides PM into five distinct categories as follows: category '0': no myopic retinal lesions, category '1': tessalated fundus, category '2': diffuse chorioretinal atrophy, category '3': patchy chorioretinal atrophy, and category '4': macular atrophy. Additional features of "plus lesions" can be assigned for lacquer cracks, choroidal neovascularization, and Fuchs' spots. MM tends to progress. Over a mean follow up of 18 years in a retrospective observational case series of 810 eyes, the authors reported progression of MM in 58.6% of all eyes, and 74.3% in eyes with pre-existing PM²⁹. Notably, META-PM is based on fundus photographs

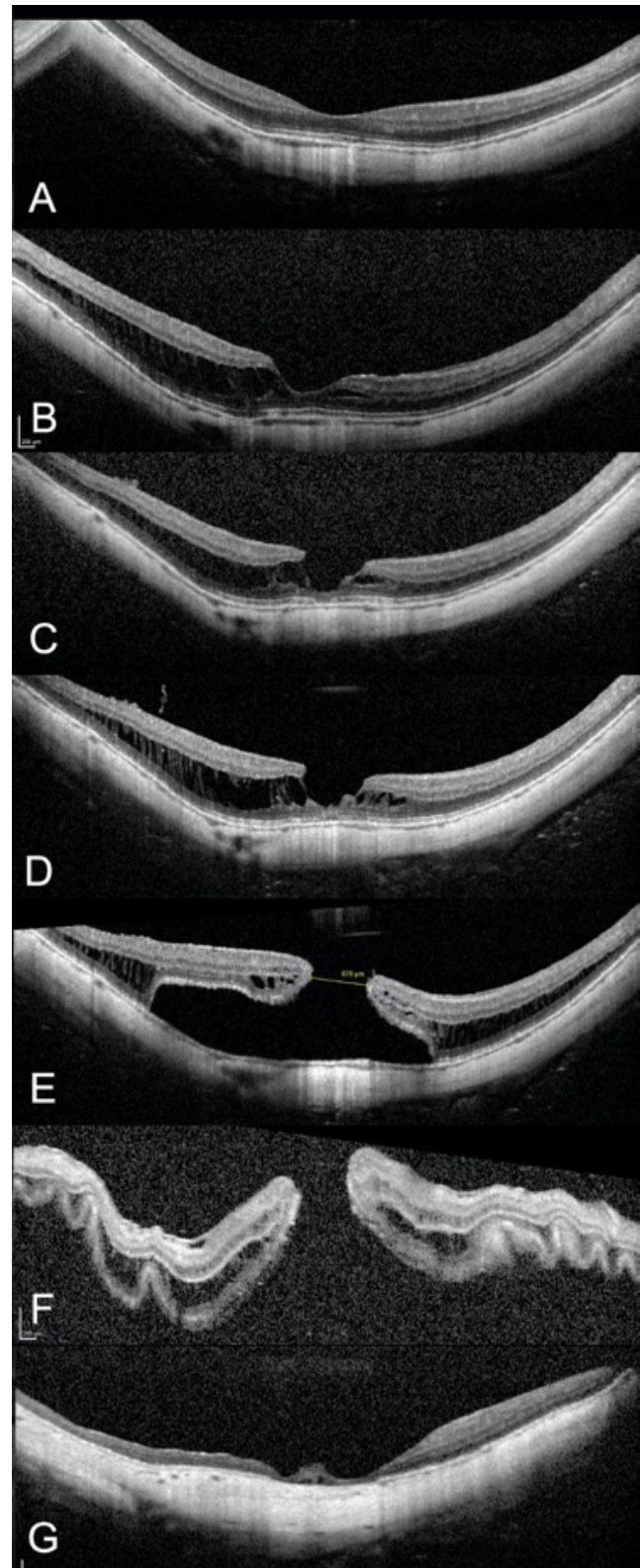


Figure 3. Progression of MTM from baseline (A) to predominantly outer retinal schisis/stage 2a at 3 years (B) followed by progressive outer retinal schisis and increasing lamellar changes/stage 2b at 4 years (C) and 5 years (D). 9 months after (D) the appearance of a macular schisis detachment/stage 3c was noted (E) with extension of the retinal detachment to outside the macula after another month (F). The patient was treatment with vitrectomy, membrane and ILM peel, and silicone oil with continued anatomical success and 20/200 vision at 2 years follow up but significant retinal atrophy (G)

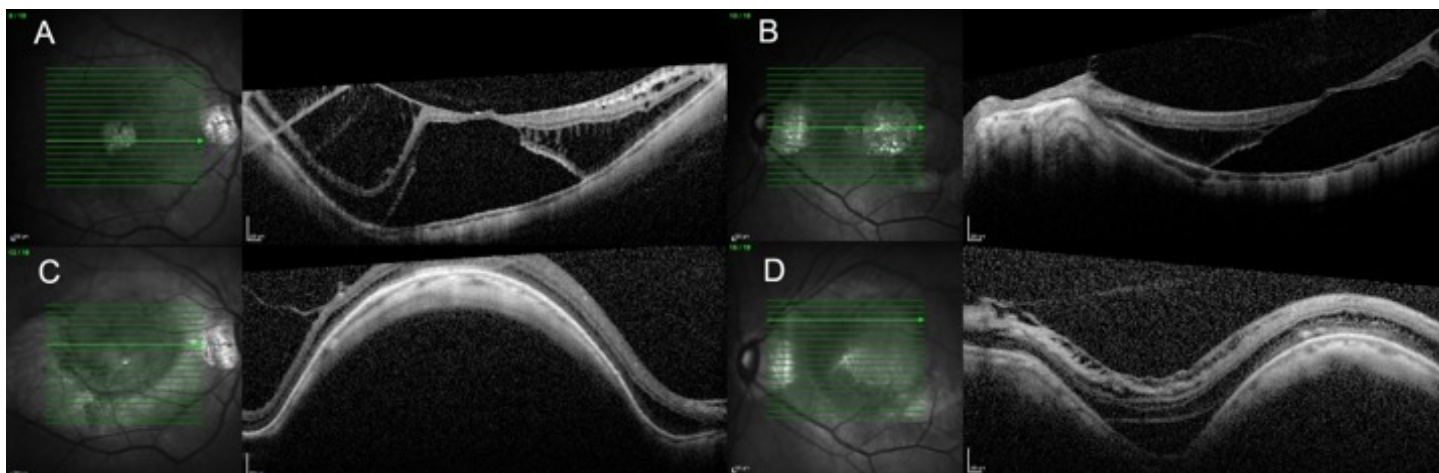


Figure 4. Pre-operative OCT of stage 4b MTM in the right (A) and left eye (B) of a -12 myope with symptomatic visual decline to 20/100 and 20/70 respectively. Post-operative OCT after successful macular buckling surgery in the right (C) and left eye (D) with reduction in retinoschisis and resolution of macular detachment. Visual acuity improved to 20/30 OU with a concurrent reduction of 5 diopters of myopia.

and does not incorporate OCT findings. Researchers have recently attempted to supplement the META-PM classification with OCT features and demonstrated that choroidal thinning is associated with the progression of myopic maculopathy. They established cut-off values of choroidal thickness of $<56.5 \mu\text{m}$ located $3000 \mu\text{m}$ nasally from the fovea to define peripapillary choroidal thinning, and $<62 \mu\text{m}$ subfoveally to define macular choroidal thinning, two terms proposed as the OCT equivalent to the subtypes of category 2 diffuse chorioretinal atrophy in META-PM. Unfortunately, to date, no established treatment for MM exists.

Conclusion

OCT has emerged as a foundational technology in the management of PM. The relative hypopigmentation of myopic fundi and microstructural changes associated with pathology means that biomicroscopy alone is insufficient in the assessment of PM eyes. Advances in OCT imaging continue to enhance our understanding of PM and it is imperative for the eyecare specialist to leverage these advantages in the care of our patients.

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