

CHAIR-SIDE REFERENCE: MYOPIC MACULOPATHY

MYOPIC MACULOPATHY

Myopic maculopathy refers to structural changes at the macula induced by high myopia (refractive error $>-6.00\text{DS}$), in which an excessive axial length and/or posterior staphyloma is the main common factor in conjunction with other factors. While several classification systems have been proposed, Ruiz-Medrano et al.¹ have suggested categorising the spectrum of myopic maculopathy into atrophic, tractional and neovascular components.

This chair-side reference depicts the various manifestations of myopic maculopathy using multi-modal imaging techniques. For peripheral myopic changes, please refer to the Centre's chair-side reference on peripheral retinal lesions.

All management recommendations below are based on the assumption that other coexisting pathologies requiring more urgent follow-up are absent.

Ref: Ruiz-Medrano J, Montero JA, Flores-Moreno I et al. Myopic maculopathy: Current status and proposal for a new classification and grading system (ATN). Prog Ret Eye Res . 2019;69:80-115.

ATROPHIC ALTERATIONS IN MYOPIC MACULOPATHY

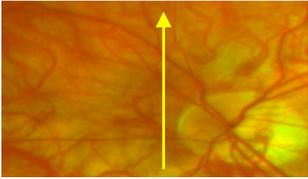
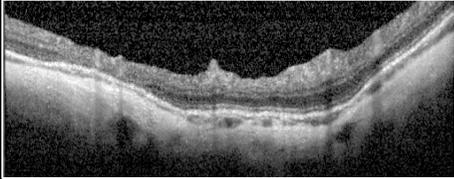
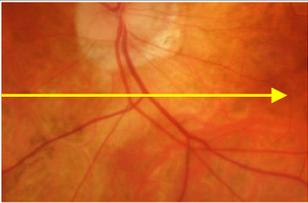
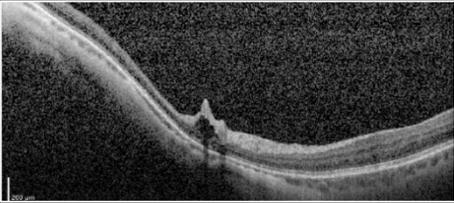
Optomap/Retinal Photo	Fundus Autofluorescence (FAF)	Optical Coherence Tomography (OCT)	Description
Tesselated Fundus			<ul style="list-style-type: none"> Increased visibility of choroidal vasculature secondary to attenuation of RPE. OCT imaging shows an intact retinal profile with no associated retinal atrophy. <i>Routine review required</i>
Diffuse Chorioretinal Atrophy			
			<ul style="list-style-type: none"> Yellowish-white appearance to the posterior pole, starting at the optic disc and macula and spreading to involve the entire staphyloma. Associated with development of choroidal neovascularisation. <i>Annual review required</i>
Multifocal (Patchy) Chorioretinal Atrophy			
			<ul style="list-style-type: none"> Well defined greyish-white areas of atrophy in the macular area and around the disc, and choroidal vessels are visible within these areas. In advanced cases, the sclera is visible within areas of atrophy. OCT imaging shows complete loss of the choriocapillaris and over time can develop to loss of the outer retina and RPE. Associated with an increased risk of developing myopic neovascularisation. <i>6-12 monthly review required</i> <i>Advise patient to perform Amsler grid self-monitoring</i> <i>Consider referral to ophthalmology to rule out CNV</i>

TRACTIONAL ALTERATIONS IN MYOPIC MACULOPATHY

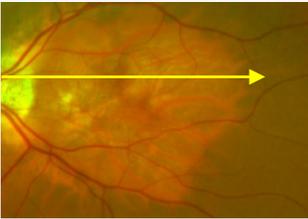
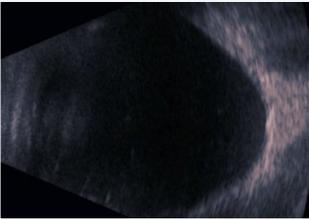
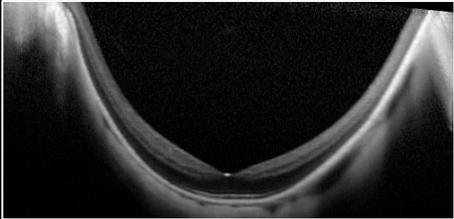
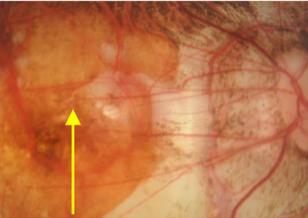
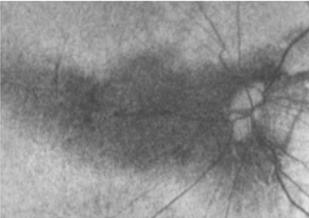
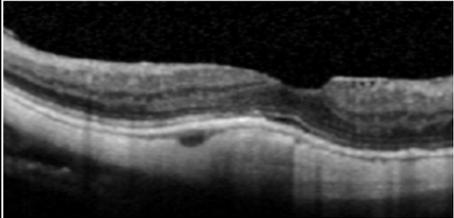
Optomap/Retinal Photo	Fundus Autofluorescence (FAF)	Optical Coherence Tomography (OCT)	Description
Myopic Macular Schisis (Myopic Foveoschisis)			<ul style="list-style-type: none"> • Separation of intraretinal layers, typically affecting the outer retinal layers. • Often no visual symptoms at onset. • Associated with axial length >31mm, chorioretinal atrophy and vitreoretinal interface disorders. • Slowly progressive, and may lead to foveal detachment or macular hole. <p><i>Annual review required</i> <i>Advise patient to perform Amsler grid self-monitoring</i> <i>Reduced VA and/or structural progression: referral is indicated</i></p>
			<ul style="list-style-type: none"> • ILM detachment (yellow arrow) has a similar appearance to an epiretinal membrane, except that there are columns that bridge the gap between the ILM and the retinal surface. • Strongly associated with myopic foveoschisis. • Retinoschisis (blue arrows) most commonly occurs in the inner and outer plexiform layers. <p><i>Annual review required</i> <i>Reduced VA and/or structural progression: referral is indicated</i></p>
Tractional inner limiting membrane (ILM) detachment and Retinoschisis			<ul style="list-style-type: none"> • Separation of the neurosensory retina from the RPE, which is typically shallow. • Associations with axial length >31mm, chorioretinal atrophy and vitreoretinal interface disorders. <p><i>Referral is indicated.</i></p>
			<ul style="list-style-type: none"> • Occurs secondary to tractional forces secondary to vitreomacular adhesion. • Risk of hole formation is higher if an ERM is seen on OCT exerting horizontal or oblique tractional force. • Absence of PVD and foveal sparing by retinoschisis are associated with increased stability. • Prognosis is poor with axial length >30mm and/or hypo-AF at the fovea. <p><i>Referral is indicated.</i></p>
Foveal Detachment			
Myopic Macular Hole			

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TRACTIONAL ALTERATIONS IN MYOPIC MACULOPATHY - Continued

Optomap/Retinal Photo	Fundus Autofluorescence (FAF)	Optical Coherence Tomography (OCT)	Description
Vascular Microfolds 	FAF not available		<ul style="list-style-type: none"> • Peaks of increased retinal thickness corresponding to retinal vasculature. • Seen in up to 44% of those with pathological myopia using OCT imaging. • In eye with paravascular cysts, the incidence of retinoschisis at the vessels is much higher than if cysts alone are present. <p><i>Routine review required</i></p>
Paravascular Cysts 	FAF not available		<ul style="list-style-type: none"> • Small hollow spaces seen on OCT imaging adjacent to large retinal vessels. • Detected in 50% of high myopes when examined with OCT imaging. • Increased occurrence with age, axial length, degree of myopia and presence of posterior staphyloma. • Associated with paravascular lamellar holes (if cysts rupture) and retinoschisis, particularly when seen with vascular microfolds. <p><i>Routine review required</i></p>

ALTERATIONS TO GLOBE MORPHOLOGY

Staphyloma 			<ul style="list-style-type: none"> • Posterior protrusion of the globe that is accompanied by a stretching of the posterior fundus. • Present in up to 90% those with high myopia and prevalence increases with age. • Clinical appearance: tessellated fundus and/or horizontal ellipse-shaped fundus pallor, typically extending from the nasal side of the disc towards the macula. • OCT imaging shows posterior bowing of the sclera, choroid and retinal layers. <p><i>Annual review required</i></p>
Dome-Shaped Macula 			<ul style="list-style-type: none"> • Convex elevation of the macula within an area of posterior staphyloma. • Most easily seen on vertical OCT line scans. • May present with reduced vision and metamorphopsia. • Associated with development of subretinal fluid, myopic choroidal neovascularisation, full-thickness macular hole formation and retinoschisis. <p><i>Annual review required. Advise patient to perform Amsler grid self-monitoring</i> <i>Reduced VA, subretinal fluid or neovascularisation: referral is indicated</i></p>

NEOVASCULAR ALTERATIONS IN MYOPIC MACULOPATHY

Optomap/Retinal Photo	Fundus Autofluorescence (FAF)	Optical Coherence Tomography (OCT)	Description
Lacquer Cracks			<ul style="list-style-type: none"> • Appear as a yellow-white lines within the macular in highly myopic eyes. • Consistent with ruptures in Bruch's membrane caused by stretching of ocular tissue with axial elongation, but prevalence not correlated with axial length. • Strong association with myopic choroidal neovascularisation which forms along the cracks. <p><i>Annual review required</i> <i>Advise patient to perform Amsler grid self-monitoring</i> <i>Reduced VA or neovascularisation: referral is indicated</i></p>
Myopic Choroidal Neovascularisation (CNV)			
			<ul style="list-style-type: none"> • Funduscopically appears as an area of grey discoloration, sometimes with a pigmented border. • Presents with decreased vision, metamorphopsia and central scotoma if located within the macula. • OCT imaging shows a hyper-reflective lesion above the level of the RPE with intra-retinal or sub-retinal fluid (hypo-reflective spaces within or beneath the retinal layers). • Fluorescein angiography may aid diagnosis. <p><i>Referral is indicated.</i></p>
Foster-Fuch's Spot			<ul style="list-style-type: none"> • A raised round or oval-shaped pigmented lesion. • Indicates proliferation of RPE pigmentation. • Often adjacent to a focal region of chorioretinal atrophy, consistent with an area of regressed choroidal neovascularisation. <p><i>Annual review required</i> <i>Advise patient to perform Amsler grid self-monitoring</i> <i>Consider referral to ophthalmology to rule out CNV</i></p>

A NOTE ON GLAUCOMA AND HIGH MYOPIA

Myopia and myopic disc configuration resulting from long axial lengths can be associated with weakening of the optic nerve head tissues with instability particularly at the lamina cribrosa, leading to increased susceptibility to insult in glaucoma.

Myopia and myopic disc configuration can also confound interpretation of the optic nerve and visual field result, mimicking glaucoma. Longitudinal data can assist in distinguishing myopic optic neuropathy from progressive and treatable glaucoma.