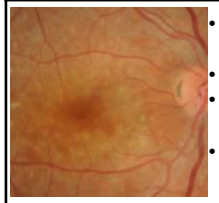
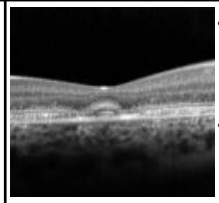
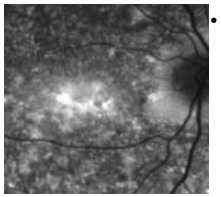
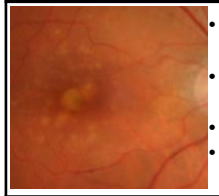
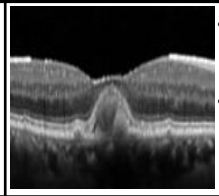
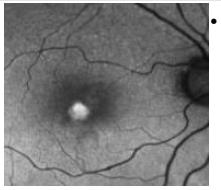
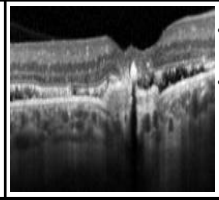
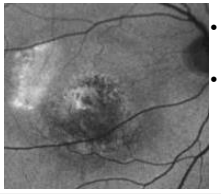
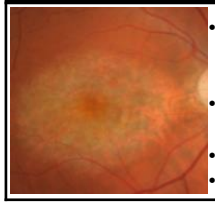
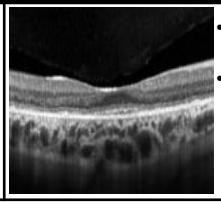
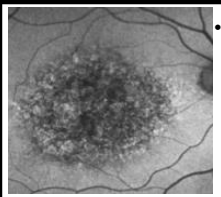
Centre for Eye Health

Chair-side Reference: Non AMD Macular Conditions

Optometry NEW SOUTH WALES AUSTRALIAN CAPITAL TERRITORY

Age related macular degeneration (AMD) is the most common pathology affecting the macula, however there are many other ocular conditions involving macula lesions that can mimic the appearance of AMD. The tables below give an overview of a number of conditions which may have a similar clinical presentation to AMD.

Aetiology of lesions at the macula can be broadly divided into three categories; changes related to dystrophies, pachychoroid spectrum and those arising from other causes. A macular dystrophy typically has a **painless bilateral onset with a progressive course**. Dystrophies may be inherited or occur through spontaneous genetic mutations. Pachychoroid spectrum refers to a group of macular diseases that manifest common choroidal features such as increased choroidal thickness and dilation of large choroidal vessels. It is thought to arise from disturbance to choroidal circulation. Other causes of macula lesions may be due to inflammation, infection, systemic causes and degenerative changes. These cases may be unilateral or bilateral but are likely to show less bilateral symmetry compared to a dystrophy.

Macula Dystrophy	Optical coherence tomography (OCT)	Fundus autofluroescence (FAF)
Stargardt Disease		
 • Foveal atrophy often surrounded by discrete yellowish round or pisciform flecks scattered throughout the fundus • Juvenile onset • Gradual and progressive visual decline ranging from 6/15-6/60 • Predominantly autosomal recessive inheritance	 • Hyper-reflective thickening of the retinal pigment epithelium (RPE) and thinning of the ellipsoid zone (EZ). • May be foveal sparing with regular EZ profile in some cases	 • Intense hyper-fluorescence of flecks surrounded by intervening hypo-fluorescence
Pattern Dystrophy		
 • Bilateral and symmetrical deposition of yellow, orange or grey material in different patterns • Common subtypes include butterfly, reticular dystrophy, or adult onset foveomacular vitelliform dystrophy • Age of onset varies from 30-50 years of age • Mild loss of vision and/or metamorphopsia; however, some may remain asymptomatic	 • Hyper-reflective lesions between the RPE and more inner retinal layers • Often disruptions of the EZ	 • Hyper-fluorescence of vitelliform lesions
Best Vitelliform Macular Dystrophy*		
 • Initially presents with a yellow, yolk like macular lesion • Progresses to atrophy in later stages • Variable age of onset ranging from 1st to 6th decade • Usually symptomatic before the age of 40 • Autosomal dominant inheritance • Risk of choroidal neovascularisation in later stages	 • Early lesions found between the RPE and sensory retina • Later stage may involve sub-retinal fluid, subretinal fibrosis and oedema	 • Variable hyper-fluorescence corresponding to vitelliform material • Hypo-fluorescence in atrophic areas
Central Areolar Choroidal Dystrophy*		
 • Initially parafoveal pigmentary RPE changes progressing to enlarged RPE atrophy and eventually confluent chorioretinal atrophy • VA deteriorates at age 30-50 years but may be asymptomatic until later. Usually causes profound vision loss • Occasionally photophobia associated • Autosomal dominant inheritance	 • Reduced retinal thickness with disruption of the EZ and outer retina • Remaining retinal layers are intact	 • Speckled pattern of hyper- and hypo-fluorescence confined to the macula region in a round / oval shape

* The specific diagnosis of dystrophies may be uncertain in the absence of electrophysiology results and genetic testing

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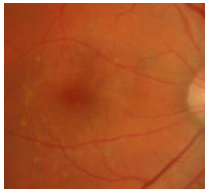
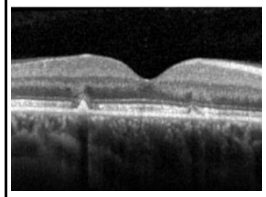
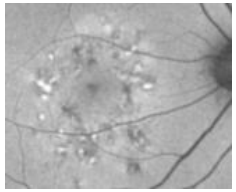
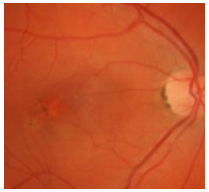
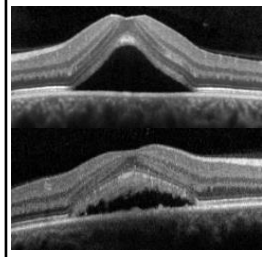
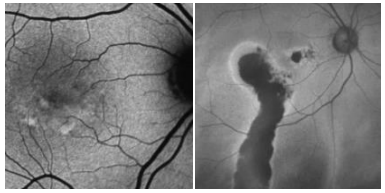
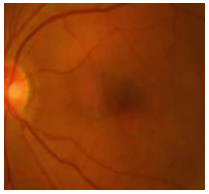
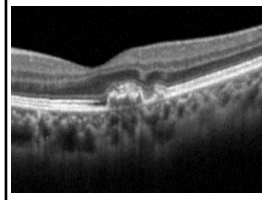
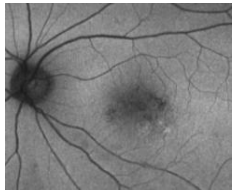
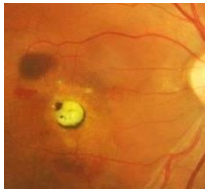
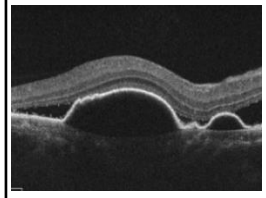
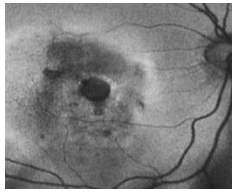
Chair-side Reference: Non-AMD Macular Conditions

Optometry NEW SOUTH WALES AUSTRALIAN CAPITAL TERRITORY

Pachychoroid Spectrum Disease	Optical coherence tomography (OCT)	Fundus autofluroescence (FAF)
Pachychoroid Pigment Epitheliopathy (PPE)		
 • Pigment alteration or minimal fundus signs • Frequently asymptomatic • Considered a forme fruste variant of CSCR • Commonly misdiagnosed as AMD or pattern dystrophy	 • Drusen like focal RPE elevation • May have serous pigment epithelial detachment (PED) • Increased choroidal thickness • No atrophy of overlying retinal layers • No present or past history of subretinal fluid	 • Granular hypo-fluoresence and/or mixed stippled hypo and hyper-fluoresence • Absence of gravitational tracts
Central Serous Chorioretinopathy (CSCR)		
 • Raised appearance of areas with residual sub-retinal fluid • Frequent pigmentary changes (hyper/hypo) • Possible unilateral blur/metamorphopsia • Most common in males age 20-50 years • Mild unilateral hyperopic shift in acute phase • Risk of CNV with increased chronicity / recurrence	 • Area of sub-retinal fluid usually with associated PED • Irregular/ thinned RPE • Chronic cases show granulation and elongation of the outer segment and subsequent outer retinal atrophy	 • Possible hyper-fluorescence associated with fluid or area of previous fluid • Hypo-fluorescent gravitational tracts if long-standing
Pachychoroid Neovasculopathy (PNV)		
 • Development of type 1 CNV following PPE and/or CSCR • Possible blur/metarmorphosia • Drusen and degenerative changes are atypical • Can progress to PCV	 • Shallow, irregular PED • With or without intraretinal or subretinal fluid	 • Mixed stippled hyper- and hypo-fluorescence
Polypoidal Choroidal Vasculopathy (PCV)		
 • Multiple and recurrent serous and haemorrhagic PEDs • Orange-red subretinal nodules • Spontaneous, recurrent subretinal and/or vitreous haemorrhage at different stages of resorption • Drusen are atypical • Typically presents as a serosanguineous maculopathy in middle-aged (50-65years) African or Asian women • Often bilateral but asymmetric	 • Haemorrhagic PED appears as a hyper-reflective elevation of the RPE with no reflectivity within or under the PED (the large, central PED pictured) • In contrast, a reflective band representing BM /choriocapillaris can be observed beneath the fluid of a serous PED (smaller PED pictured) • EDI can enhance visualisation of polyps	 • Confluent hypo-fluorescence at the sites of polypoidal lesions and granular hypo-fluorescence at the branching choroidal vascular networks • Presence of polyps best confirmed by indocyanne green angiography

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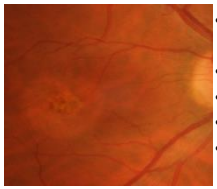
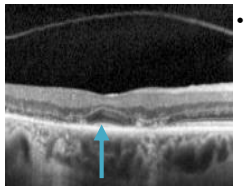
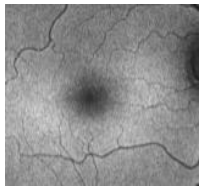
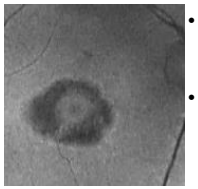
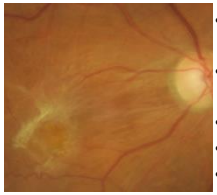
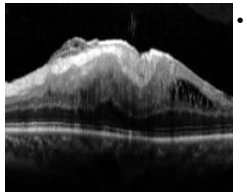
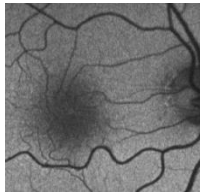
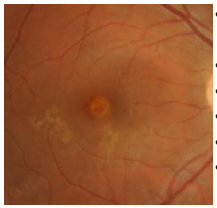
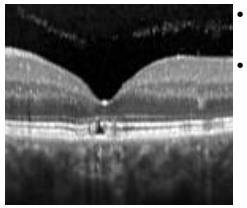
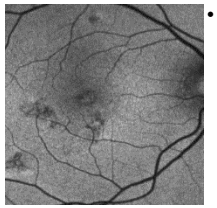
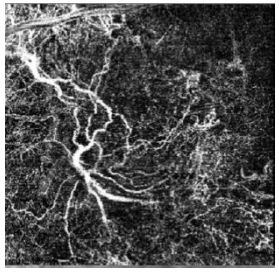
Optometry NEW SOUTH WALES AUSTRALIAN CAPITAL TERRITORY

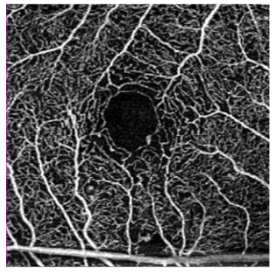
Non-AMD Condition	Optical coherence tomography (OCT)	Fundus autofluroescence (FAF)
Plaquenil Retinopathy		
 • Bilateral granular depigmentation of RPE in the macular region progressing to atrophic bull's eye maculopathy • Often asymptomatic • Possible colour vision deficit, blur and/or metamorphopsia • Medical history of Plaquenil/hydroxychloroquine • Increased risk with duration and cumulative dose (>1000mg or 5mg/kg/day)	 • "Flying saucer" sign (blue arrow) due to loss of the foveal depression and perifoveal outer retinal thinning with preservation of outer retinal structures in the central fovea	  • Fig 1: Early Disease features a ring of hyper-fluorescence • Fig 2: Late disease features a ring of hypo-fluorescence
Epiretinal Membrane/Macular Pucker		
 • Irregular light reflex or sheen which becomes more obvious with thickened membrane • May cause distortion of blood vessels, retinal wrinkling and white striae • Ranges from asymptomatic to blur/metamorphopsia • More common over age 50 • Normally follows a PVD or peripheral retinal change	 • Highly reflective layer overlying inner retinal surface with associated tractional changes (retinal thickening and pseudocystic spaces)	 • Normally typical background autofluorescence as the RPE is unaffected • Retinal dragging and distortion of blood vessels may be accentuated due to contrast
Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE)		
 • Multifocal yellow-white lesions predominantly in the posterior pole which regress over 1-2 weeks • Pigmentary changes may persist following resolution • Acute onset between 20 to 50 years of age. Idiopathic • Metamorphopsia and paracentral scotoma • Unilateral progressing to bilateral (days/weeks) • Good visual prognosis unless foveal involvement, recurrence or older age of presentation	 • Disruption of the EZ and outer retina • RPE atrophy	 • Hypo-fluorescent lesions at presentation followed by hyper-fluorescence as lesions regress

OCT angiography and macular disease

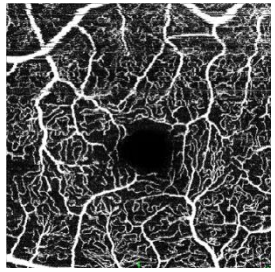
OCT angiography is a novel imaging technique that allows visualisation of retinal and choroidal vasculature without the need for dye injections. It has shown potential in diagnosing choroidal neovascularisation, visualising retinal vascular abnormalities; as well as providing new insights into macular diseases such as macular telangiectasia.



Choroidal neovascularisation in **AMD**



Microaneurysms and capillary dropout in **Diabetic Retinopathy**



Capillary rarefaction in **Macular Telangiectasia**