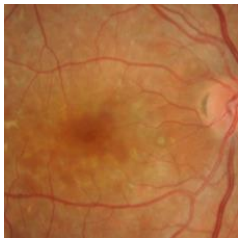
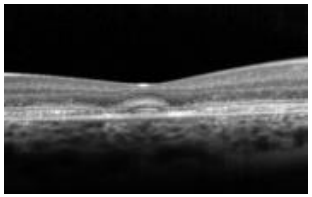
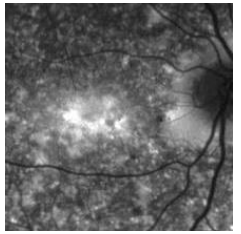
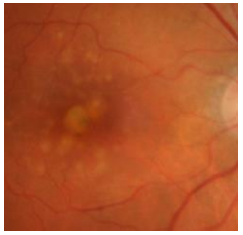
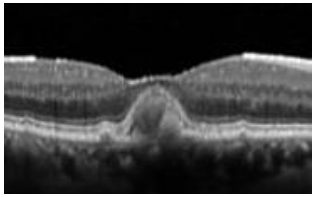
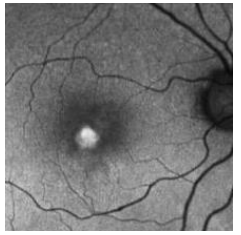
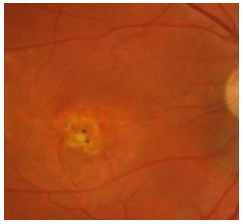
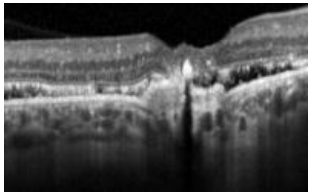
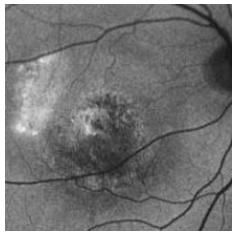
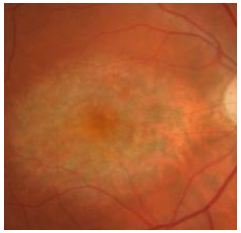
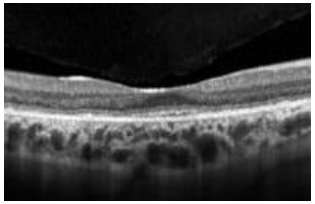
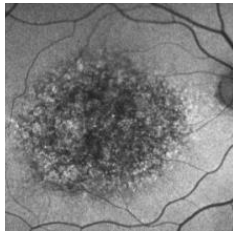
Centre for Eye Health

Chair-side Reference: Non AMD Macular Conditions

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 are commonly confused with AMD.

Aetiology of lesions at the macula can be broadly divided into two categories; changes related to dystrophies and those arising from other causes. A macula dystrophy typically has a **painless bilateral onset with a progressive course**. Dystrophies may be inherited or occur through spontaneous genetic mutations. In the case of dominant inheritance, there will likely be a clear family history however in the case of recessive or x-linked inheritance, there may be few or no family members affected. Conversely, macula problems which are not dystrophic in origin may result from a combination of factors including genetic predisposition, inflammation, infection, medical history 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 autofluorescence (FAF)
Stargardt Disease		
 • Foveal atrophy surrounded by discrete yellowish round or pisciform flecks scattered through the fundus • Juvenile onset • Gradual and progressive visual decline ranging from 6/15-6/60 • Predominantly autosomal recessive inheritance	 • Hyper-reflective thickening of the RPE layer and thinning of the ISe line • May be foveal sparing with regular ISe profile in some cases	 • Intense hyper-fluorescence of flecks surrounded by hypo-fluorescence
Pattern Dystrophy		
 • Bilateral and symmetrical deposition of yellow, orange or grey materials in different patterns • Presentations include butterfly, reticular dystrophy, 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 • Autosomal dominant inheritance	 • Hyper-reflective lesions between the RPE and more inner retinal layers. • Often disruptions of the ISe zone	 • Hyper-fluorescence of vitelliform lesions
Best Vitelliform Macular Dystrophy		
 • Initially presents with yellow, yolk like macular lesion • Progresses to geographic 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 scars 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 geographic atrophy • VA deteriorates at age 30-50 but some cases are asymptomatic until later • Occasionally photophobia associated • Autosomal dominant inheritance	 • Reduced retinal thickness with disruption of the ISe zone 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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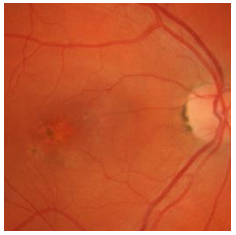
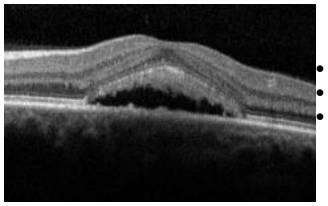
Chair-side Reference: Non AMD Macular Conditions

Non-Dystrophic Macular Changes

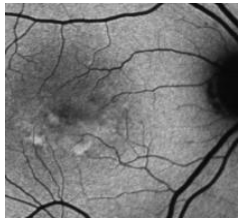
Chronic / Recurrent Central Serous Chorioretinopathy

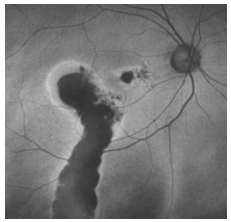


- Raised appearance of areas with residual sub-retinal fluid
- Frequently pigmentary changes (hyper/hypo)
- Possible blur/metamorphopsia unilaterally
- Most common in males age 20-50 years
- Mild unilateral hyperopic shift in acute phase
- Risk of CNV with increased chronicity / recurrence



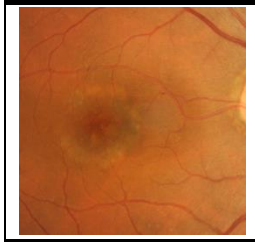
- Area of sub-retinal fluid
- Irregular/ thinned RPE
- Granulation and focal thinning of the posterior surface of the detached retina



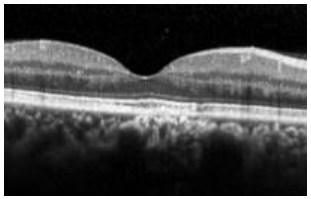


- Possible hyper-fluorescence associated with fluid or area of previous fluid
- Gravitational tracts if long-standing

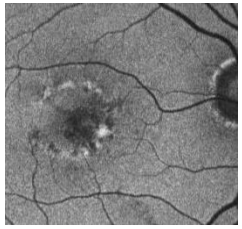
Resolved Central Serous Chorioretinopathy



- Pigmentary changes frequent (hyper/hypo) following resolution of condition
- Possible blur/metamorphopsia unilaterally
- May be recurrent
- Increased risk of CNV with increased retinal disturbance

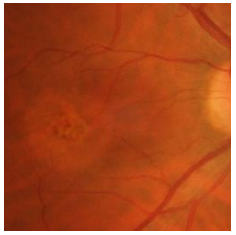


- Irregular RPE/ISe zone upon resolution of fluid

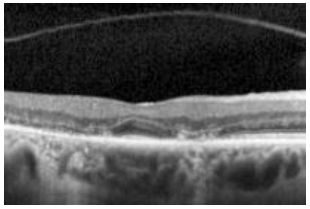


- Possible hyper-fluorescence associated with area of previous fluid
- Hypo-fluorescence if atrophic

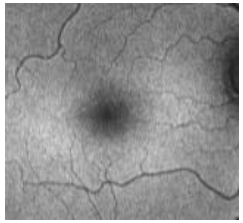
Plaquenil Retinopathy

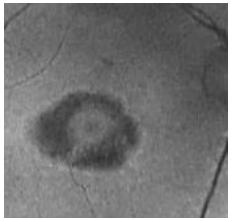


- Bilateral granular depigmentation of RPE in the macular region progressing to atrophic bull's eye maculopathy
- Often asymptomatic
- Possible colour vision deficit, blur and/or metamorphopsia
- Medical history of Plaquenil/hydroxychloroquine
- Increased risk with cumulative dose (dosage and duration)



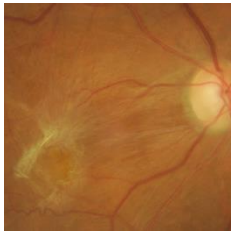
- Atrophy of outer retina in a bull's eye pattern sparing a foveal island
- Underlying increased choroidal reflectivity



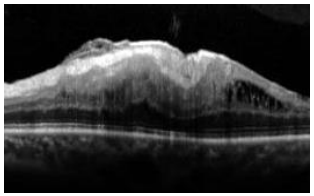


- Fig 1: Early ring of hyper-fluorescence
- Fig 2: Late ring of hypo-fluorescence

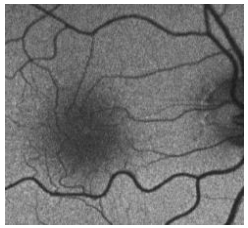
Epiretinal Membrane/Macular Pucker



- 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

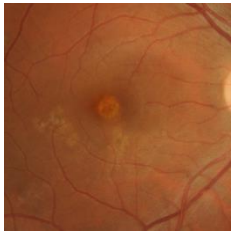


- Highly reflective layer overlying inner retinal surface with underlying tractional changes (retinal thickening and pseudocystic spaces)

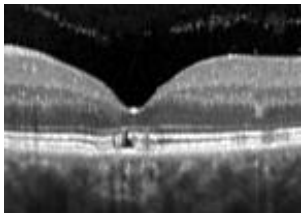


- Normally typical background autofluorescence as the RPE is unaffected
- Distortion of blood vessels may be accentuated

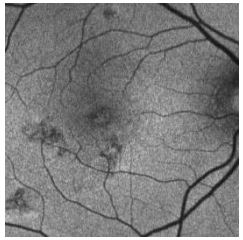
Acute Posterior / Multifocal Placoid Pigment Epitheliopathy (APMPPE)



- Multifocal yellow-white lesions predominantly in the posterior pole which regress over 1-2 weeks
- Pigmentary changes may persist following resolution of white dots
- Acute onset between 20 to 50 years of age
- Metamorphopsia and paracentral scotoma
- Unilateral progressing to bilateral (days/weeks)
- Idiopathic
- Good visual prognosis unless foveal involvement, recurrence or older age of presentation



- Disruption of the ISe line and outer retina
- RPE atrophy



- Hypo-fluorescent lesions at presentation followed by hyper-fluorescence as lesions regress