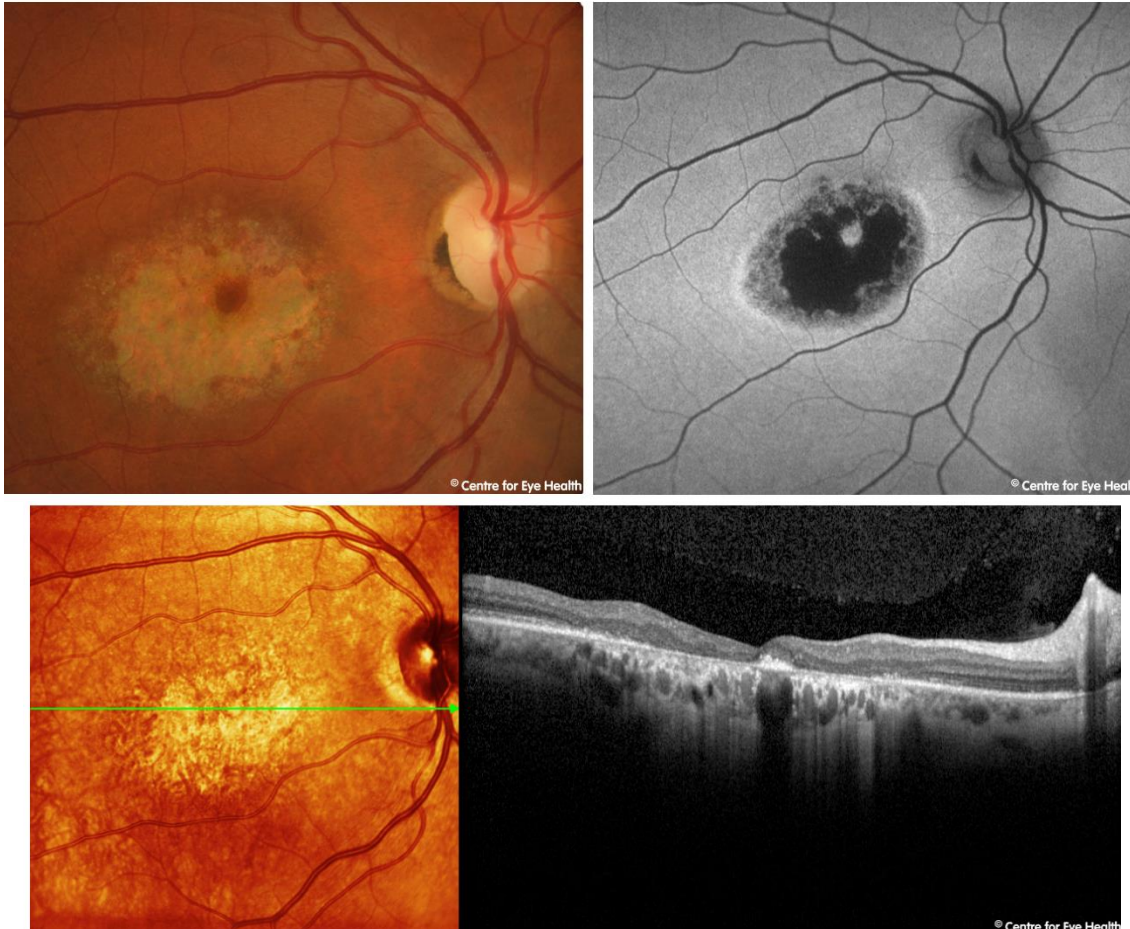




CFEH Facebook Case #89

A 45 year old Middle Eastern male presents complaining of blurry vision that has progressively worsened over the previous 4-5 years. He mentions a colour vision deficiency that was diagnosed as a child and testing with the D-15 reveals a protan defect. Best corrected acuities are 6/48 in each eye. Both eyes have a similar appearance so only the right eye is shown below. What are your differential diagnoses and what further testing could be performed to help differentiate them?



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ANSWER

Retinal images show a patchy bilateral retinal atrophy around the maculae with a ring-shaped appearance and a small central region of relative sparing. OCT shows a distortion of the foveal pit and disorganisation of the retinal layers. There is loss of the outer retinal layers (the outer nuclear layer, inner segment ellipsoid layer, external limiting membrane and RPE). There is also a hyper-reflective sub-retinal elevation with posterior shadowing. These findings are consistent with a diagnosis of either central areolar choroidal dystrophy (CACD), although a diagnosis of cone dystrophy must also be excluded.

CACD is an inherited condition characterised by an area of photoreceptor, RPE and choriocapillaris atrophy at the central macula. Patients typically present with reduced vision in their 40-50's. Progressive atrophy eventually affects the fovea around the age of 70, causing significant central vision loss. Clinically, OCT shows loss of the outer retinal layers, such as in this patient and a ring of hyper-autofluorescence in early stages, followed by an area of marked hypo-autofluorescence in the later stages.

Electrophysiology testing (below) confirmed a diagnosis of macular dystrophy. For further information on the clinical uses and basic interpretation of electrophysiology you can register [here](#) to attend our February webinar "Electrophysiology 101 – what every optometrist needs to know", presented by Michael Yapp and Dr Nagi Assaad on Tuesday 13th February 2018 at 6:30pm.

