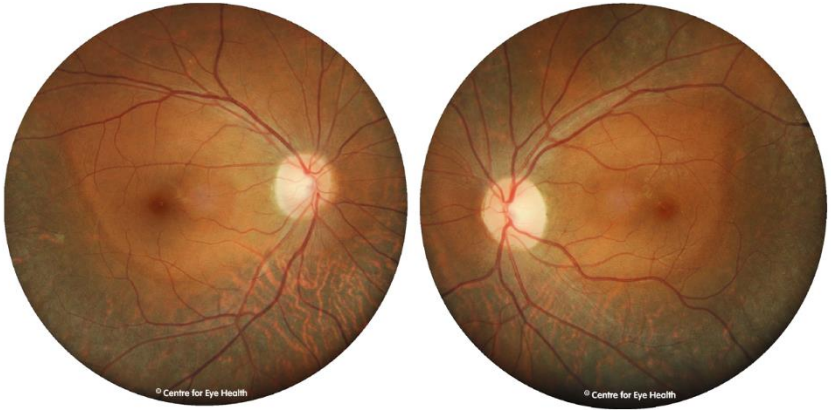
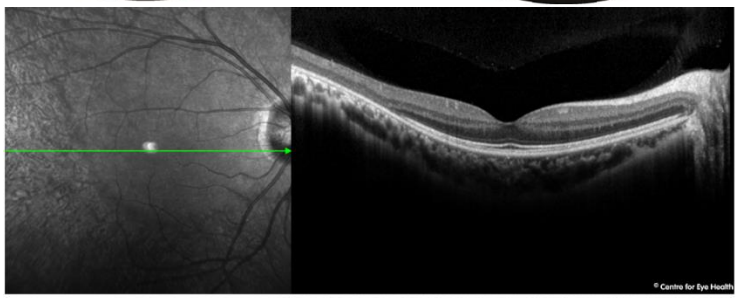
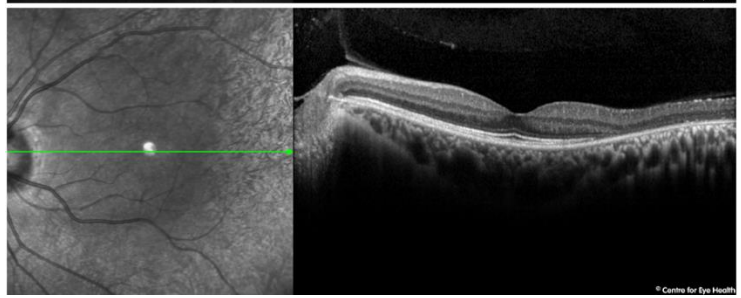
CFEH facebook Cases

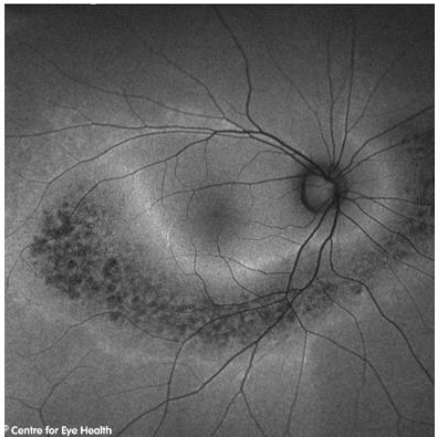
CFEH Facebook Case #53

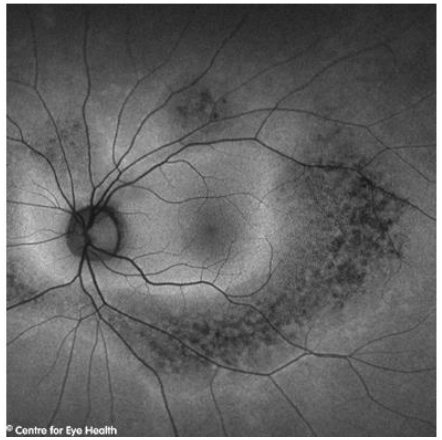
A 35 year old asymptomatic Indian male was referred for a retinal assessment, suspected to have retinitis pigmentosa (RP). Acuities were 6/3.8- OD and 6/4.8 OS. Colour vision testing with the L'Anthony desaturated D-15 was passed in the right eye and failed in the left with 2 crossings along a tritan axis. He reported a history of cat scratch disease and associated vision loss in the left eye 16 years ago but otherwise no significant family, medical or ocular history. He doesn't have RP, but what retinal condition does he have?











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ANSWER

Posterior polar annular choroidal dystrophy (PPACD)

PPACD is a rare condition characterised by progressive chorioretinal atrophy around the vascular arcades and optic disc. There is typically some preservation of the choriocapillaris at the junction between the area of atrophy and normal which appears as a fringe of hyper-autofluorescence. The OCT images show attenuation of the outer retinal layers with thinning of the outer nuclear layer and an absent IsE zone.

Electrophysiology testing showed findings consistent with PPACD. A full field ERG showed the photopic responses (cone responses) to have slightly delayed b wave of reduced amplitude in both eyes. Pure rod responses were substantially reduced in amplitude with prolonged a and b waves (see image below).