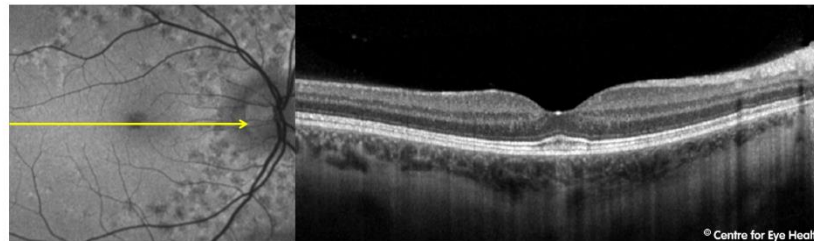
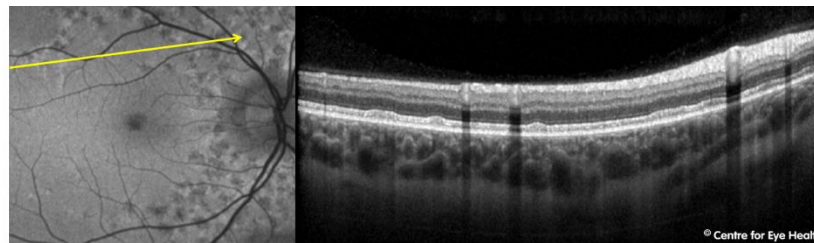
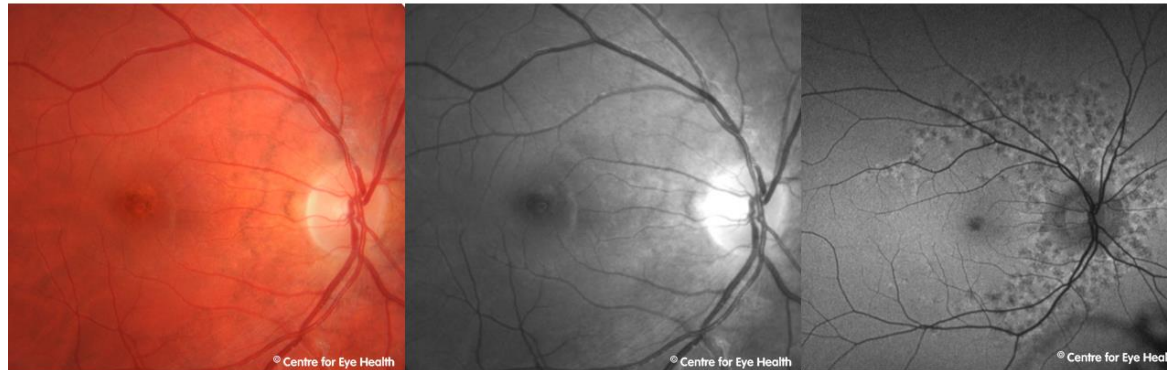




## CFEH Facebook Case #131

A 22 year old female presented for a macular assessment. Her acuities were 6/6 in each eye with no distortion on Amsler grid and an unremarkable ocular and medical history. Both eyes had a similar appearance, so only the right eye is shown here. What would be the most likely diagnosis for this patient?



# Answer

In this patient's retinal photo, subtle darkly pigmented lines may be seen around the posterior pole. These extend from the peripapillary region out to the superior and inferior vascular arcades. There are numerous areas of hypo-autofluorescence seen along both vascular arcades, and on OCT areas of focal RPE thickening can be seen. This presentation is consistent with a diagnosis of reticular dystrophy of the RPE which is a type of pattern dystrophy.

Early presentation of this condition is characterised by pigmentary changes which start at the fovea and move outwards from there. This pigment eventually fades, leaving RPE atrophy in its place. Fortunately vision is usually minimally affected, even in advanced cases, however in some cases atrophy and choroidal neovascularisation can cause vision loss.

Associated ocular conditions may include spherophakia with myopia and luxated lens, partial iris atrophy, scleral staphyloma, convergent strabismus, and choroidal neovascularisation. There may also be systemic associations, including deaf-mutism and choreatiform behaviour (involuntary movement disorder).

An annual review is recommended, and family members should also be screened as this condition is hereditary.