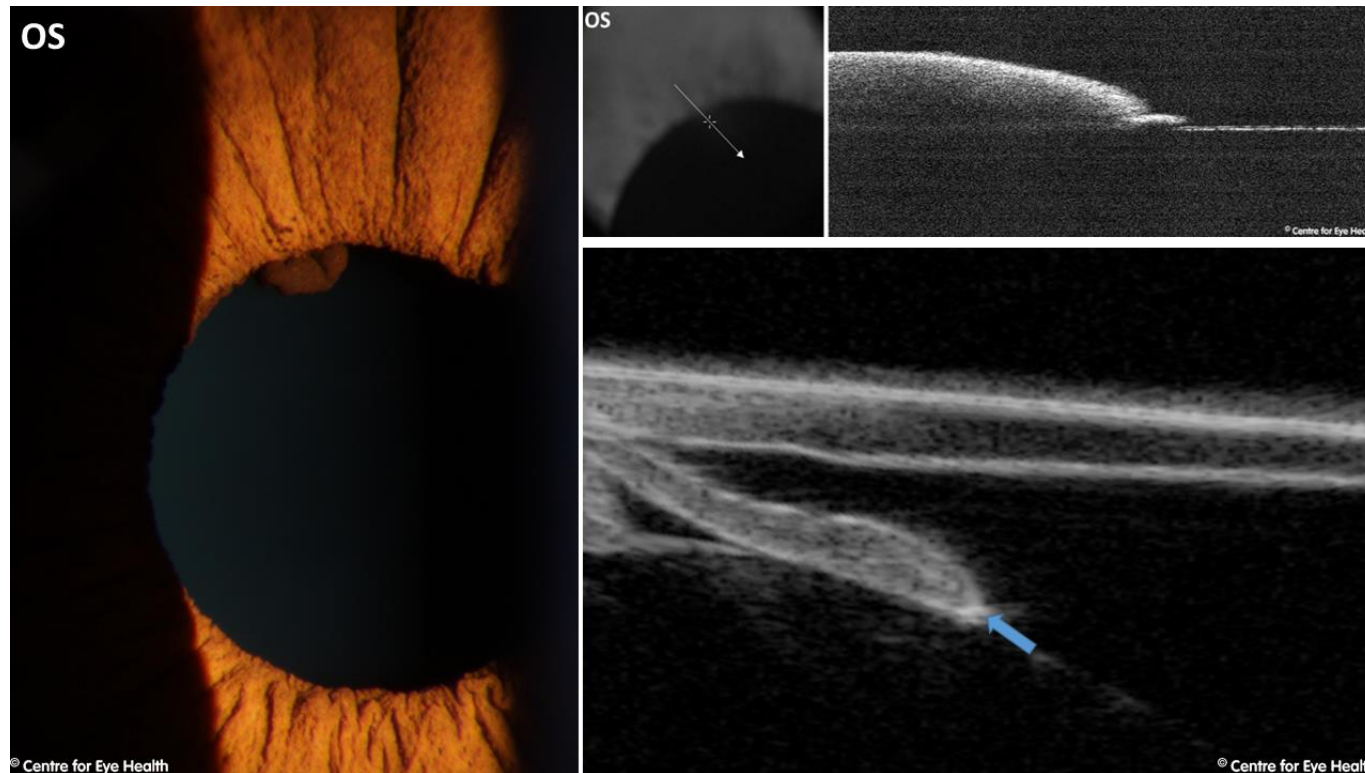




## CFEH Facebook Case #129

A 23 year old male was referred for further investigation of pigmented iris lesions found at the pupil margin in each eye (For the purpose of this case, only the left eye is shown). Pupils were equal, round and reactive to light and accommodation in both eyes and vision was 6/6 OU. What is the abnormality and what are the possible causes of such a finding?



# Answer

Anterior eye imaging revealed hypertrophy of the iris pigment epithelium across the anterior iris surface, nasally in the right eye and superiorly in the left. Optovue OCT and UBM scans showed increased hyper-reflectivity of these lesions. They appeared to arise from the posterior margin of the iris, with no associated mass of cystic changes posterior to the lesions.

This appearance is consistent with a diagnosis of mild ectropion uveae (EU) which is defined as the presence of iris pigment epithelium on the anterior iris surface.

EU can be either congenital or acquired. Acquired cases may be caused by iris neovascularisation / neovascular glaucoma, or they may be secondary to inflammatory, ischemic, or neoplastic conditions affecting the iris. Acquired EU are usually progressive due to membranous iris traction.

Congenital cases of EU are rare and non-progressive but are often associated with systemic conditions including (most commonly) neurofibromatosis 1, but also Prader-Willi syndrome, Rieger anomaly, and facial hemihypertrophy/dysgenesis. Congenital EU is associated with anterior insertion of the iris, dysgenesis of the drainage angle, and has a high rate of glaucoma (up to 90% of cases develop glaucoma) so these patients should be monitored carefully.