

# Condition Spotlight:

## Pachychoroid Spectrum Disease

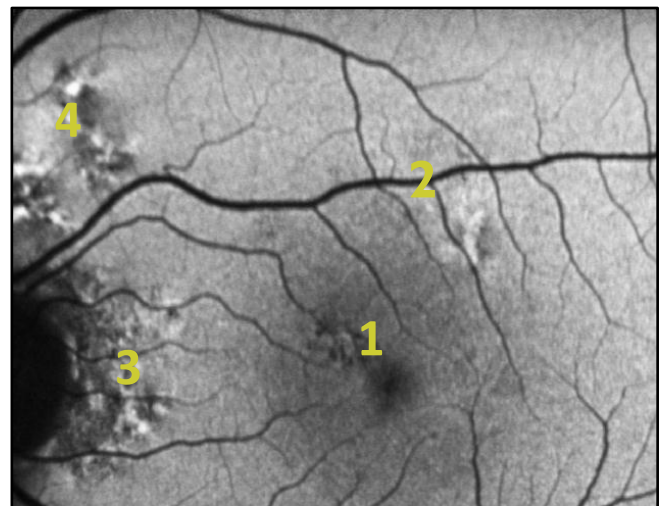
### Case report by Rebecca Tobias

A 47 year old Caucasian male was referred to CFEH following findings of RPE changes at the macula. Ocular history included right strabismus surgery but was otherwise unremarkable. Medical history includes sleep apnoea and hypertension and current medications include Seretide, Accupril and Lopressor.

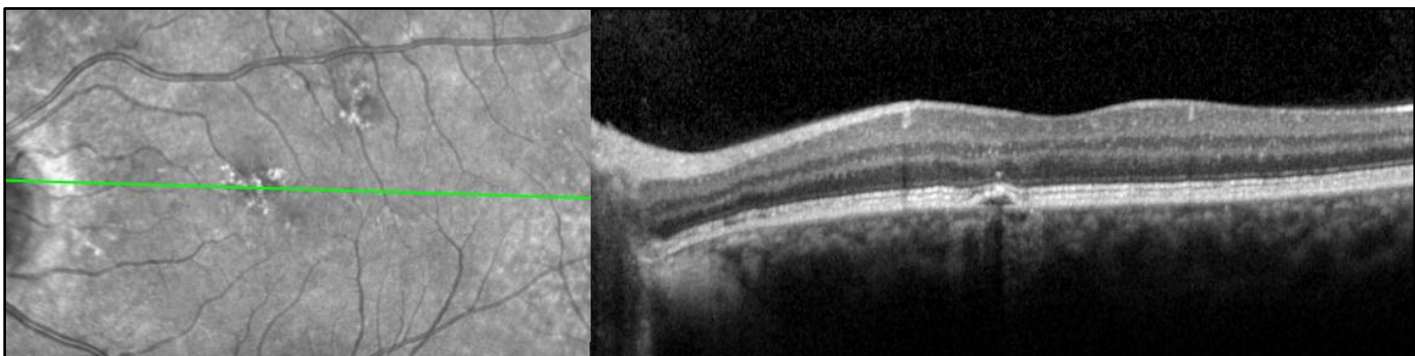
	OD	OS
<b>Entering aided acuities</b>	6/19-2 (NIPH) – reduced due to amblyopia and strabismus	6/7.5
<b>Amsler grid</b>	Unremarkable	Unremarkable
<b>Contrast sensitivity – MARS test (normal Range 1.72 – 1.92)</b>	1.68 units	1.68 units



Funduscopy revealed areas of mottled RPE changes in both eyes. The left eye (above) showed 3 areas of RPE changes within and around the macula (1,2) and two near the optic nerve (3,4).



Fundus Autofluorescence (FAF) showed mixed stippled hyper- and hypo- autofluorescence corresponding to the four areas of RPE changes seen on fundus photography. Note that these changes are more evident using fundus autofluorescence than with funduscopy.



OCT imaging showed an isolated shallow PED and disruption of the ISe zone. There is just visible overlying pigment migration at this point, seen as a hyperreflective foci on the OCT scan. There is also mottling of the ISe zone corresponding to the changes adjacent to the disc.

**Summary:** The findings were consistent with pachychoroid pigment epitheliopathy (PPE), part of a new classification of retinal disorders associated with pachychoroid (thickened choroid) related vascular changes.

**Management plan:** Review in 6 months with repeat imaging. The patient was advised to self-monitor with an Amsler grid in the interim and report sooner should any changes be noted.



## Clinical implications

- ***Pachychoroid pigment epitheliopathy (PPE) forms part of a spectrum of retinal disorders known as pachychoroid spectrum disease.***

Pachychoroid spectrum disease refers to conditions with common features of increased choroidal thickness, reduced fundus tessellation, drusenoid RPE changes, areas of hyper and hypo autofluorescence that are in excess of RPE changes noted clinically, and the presence of small PEDs overlying areas of thickened choroid (Warrow, 2013) (Dansingani K. B., 2015). Recently it has been proposed that the characteristically increased choroidal thickness results from pathologically dilated choroidal vessels, termed “pachyvessels” (Dansingani K. B., 2015). These changes are best observed with swept source OCT or enhanced depth imaging on spectral domain OCT. In addition to PPE, other conditions included in the spectrum include central serous chorioretinopathy (CSCR), polypoidal choroidal vasculopathy (PCV) and pachychoroid neovasculopathy. PPE is likely to be a “forme fruste” manifestation of CSCR (Warrow, 2013) as it involves no history of serous macular detachment or sub-retinal fluid but other clinical characteristics are similar.

- ***Pachychoroid spectrum disease is associated with choroidal neovascularisation (CNV).***

Choroidal neovascularisation is to the growth of new vessels from the choroid which break through Bruch’s membrane. The sub-division of choroidal neovascularisation into occult (type 1) and classic (type 2) was originally based on the patterns seen in fluorescein angiography. Classic membranes are associated with a well demarcated early hyperfluorescent area, increasing in intensity and extent by mid to late stage frames. In contrast, occult lesions are presumed when late granular choroidal leakage is noted with no discernible classic membrane pattern. These characteristics are attributed to the differing locations and penetrations of the membranes where classic lesions penetrate the RPE and therefore lie anteriorly whereas occult lesions are sub RPE (Lim, 2012). A greater understanding of these and other characteristics has allowed improved detection and potential diagnosis with advancing OCT technology including OCT angiography (Jia, 2014). Studies have shown variable associations between CSCR and CNV, with the classic type (type 2) more commonly reported and occult (type 1) CNV also described. In 2013 Warrow et al. proposed that PPE patients may also develop type 1 sub-RPE neovascularisation (“pachychoroid neovasculopathy”) (Warrow, 2013) which may in turn progress to PCV (Pang, 2015). Previously this presentation may have been misdiagnosed as AMD, inflammatory chorioretinopathy or idiopathic neovascularisation.

- ***In eyes with pachychoroid spectrum disease, the finding of shallow irregular PEDs on OCT may be a significant indicator of the development of type 1 neovascularisation.***

A recent study documented neovascular networks in 95% of patients with coexisting features of shallow irregular PEDs and pachychoroid characteristics (as detected by OCT angiography years after the initial diagnosis). In most cases, scans through the PEDs showed networks of ‘trunk vessels’ with identifiable feeder vessels. (Dansingani K. B., 2015). In pachychoroid neovasculopathy, type 1 neovascularisation is typically found where the pachyvessels are adjacent to the outer aspect of the RPE-Bruch’s membrane complex (Pang, 2015) (Dansingani K. B., 2015).

## References

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