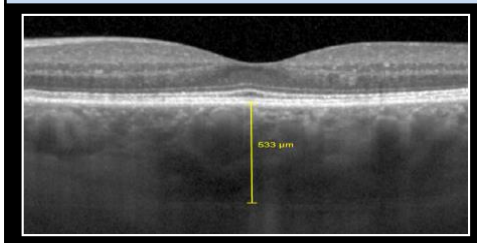
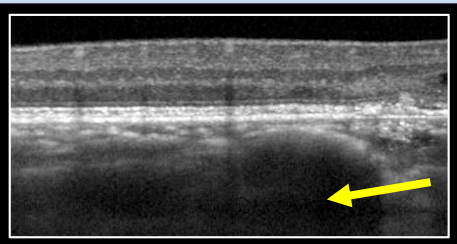
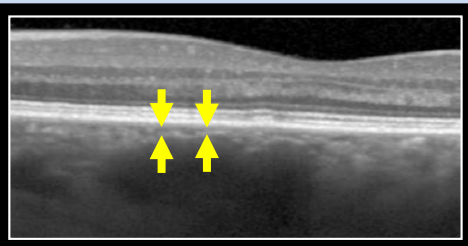
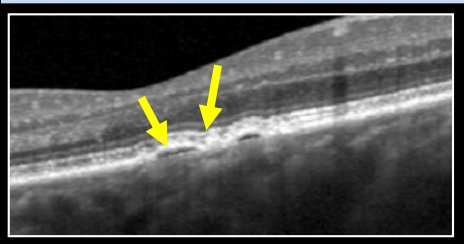
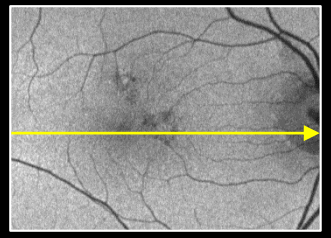
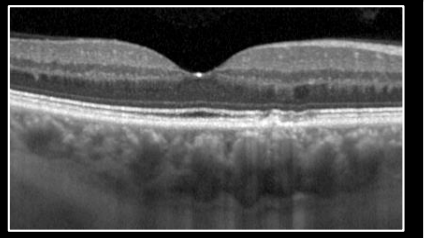
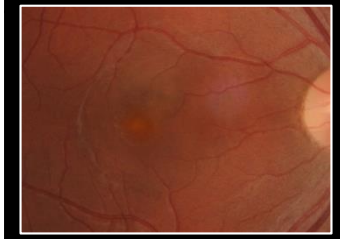
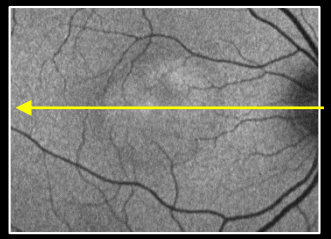
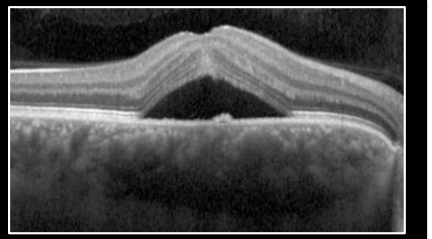
Centre for Eye Health

CHAIR-SIDE REFERENCE: PACHYCHOROID DISEASE SPECTRUM

PACHYCHOROID DISEASE SPECTRUM

The Pachychoroid disease spectrum (PDS) refers to a group of macular conditions characterised by a thickened ("pachy") choroid. These conditions are linked to both structural and functional alterations in the choroid, with more recent associations involving vortex vein congestion and scleral abnormalities. The shared features of these conditions are illustrated below:

Increased choroidal thickness	Dilated large choroidal vessels	Overlying choriocapillaris attenuation	Associated RPE disturbances
			

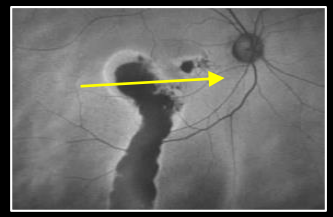
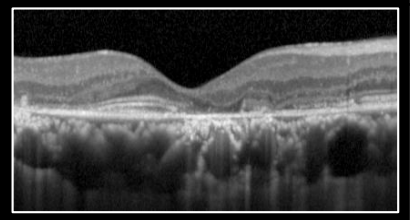
Retinal photo	Fundus Autofluorescence	OCT	Description
Pachychoroid Pigment Epitheliopathy (PPE)			
			• **Definition:** considered a forme fruste variant of central serous chorioretinopathy (CSCR), with no history of current or past sub-retinal fluid. • **Symptoms:** typically asymptomatic. • **Fundus exam:** can have minimal signs, though pigment alterations may be present. • **FAF:** shows granular hypo-fluorescence and/or mixed stippled hypo and hyper-fluorescence. • **OCT:** shows drusen-like focal RPE elevations, possibly with an associated serous pigment epithelial detachment (PED) *May progress to other forms of pachychoroid disease spectrum, yearly review with OCT recommended.*
Acute Central Serous Chorioretinopathy (CSCR)			
			• **Definition:** localised serous detachment of macula. • **Symptoms:** unilateral blur or metamorphospsia with a mild hyperopic shift. • **Fundus exam:** raised appearance of the macula with frequent hyper or hypo pigment changes. • **FAF** may show no apparent abnormality or hyper-fluorescence associated with the areas of sub-retinal fluid. • **OCT** shows a well-defined serous retinal detachment usually associated with a PED. • **Hi**gh proportion (78-84%) of cases self-resolve by 6 months. *Remove or modify risk factors if possible, review with repeat OCT in 3 months. If there is no improvement by 3 months and/or the patient requires optimised vision and faster recovery, consider referral to Ophthalmologist.*

Centre for Eye Health

CHAIR-SIDE REFERENCE: PACHYCHOROID DISEASE SPECTRUM

Optomap image	Fundus Autofluorescence	OCT	Description

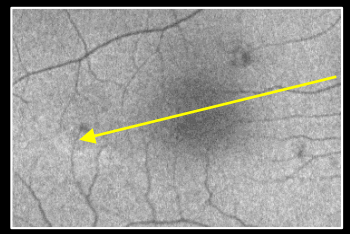
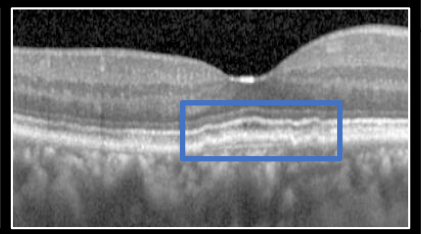
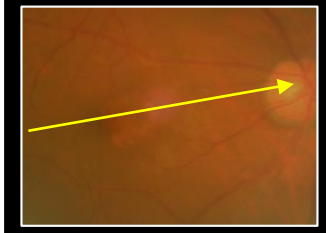
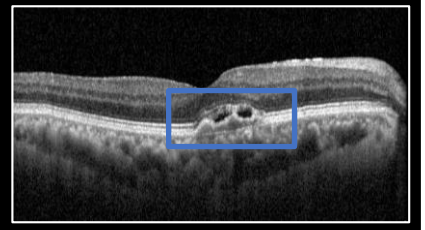
Chronic Central Serous Chorioretinopathy (CSCR)

- **Definition**: Variable in literature, but CSCR of >3-6month duration. Can be associated with widespread RPE decompensation with or without SRF.
- **Symptoms**: Reduced vision or distorted vision can occur with macular atrophy and/or MNV.
- **FAF:** Long-standing cases associated with hypo-fluorescent gravitational tracts.
- **OCT:** Shows outer retina and RPE atrophy.
- Risk of macular neovascularisation (MNV) increases with increased recurrence or chronicity.

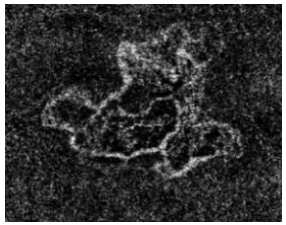
Review every 4-6 months with referral to Ophthalmologist if there is persistent subretinal fluid (SRF), vision impairment, or suspected MNV.

Pachychoroid Neovasculopathy (PNV)





FAF not available


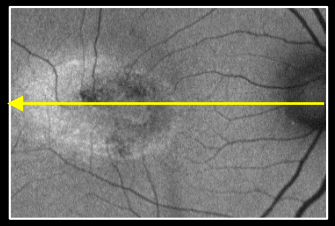
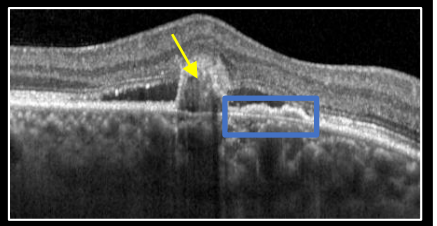
- **Definition:** Development of type 1 (sub-RPE) MNV following PPE/CSCR. Occurs on a spectrum with PCV.
- **Symptoms:** Blur/metamorphopsia, especially if exudative.
- **Fundus exam:** Varies from subtle pigmentary changes to subretinal haemorrhage depending on presence of exudation. Typical AMD drusen absent.
- **FAF** highlights RPE changes overlying thick choroid
- **OCT** shows a flat, irregular PED (double-layer sign)
- Can be non-exudative (top images) or exudative (lower images). Approximately 9% of non-exudative cases have exudative changes at 12 month review.

Refer to Ophthalmologist in the presence of exudative signs (reduced vision, fluid on OCT)



OCT angiography shows 'tangled network' of flow signal corresponding to type 1 MNV

Polypoidal Choroidal Vasculopathy (PCV)/Aneurysmal Type 1 Neovascularization

- **Definition**: Variant of PNV, features type 1 MNV and aneurysmal dilations, the latter has polyp-like structure. Associated with serous neurosensory detachment and/or submacular haemorrhage.
- **Symptoms**: blur or metamorphopsia, especially if exudative.
- **Fundus exam:** shows orange-red subretinal nodules.
- **FAF** shows ring-shaped abnormalities with hypo-autofluorescent centre (correspond to polyps) and hyper-autofluorescent surroundings.
- **OCT** shows a sharp PED peak (yellow arrow) and surrounding flat, irregular PED (blue box).
- Indocyanine green angiography is the gold standard for diagnosing PCV.

Refer to Ophthalmologist.

Centre for Eye Health

CHAIR-SIDE REFERENCE: PACHYCHOROID DISEASE SPECTRUM

Optomap image	Fundus Autofluorescence	OCT	Description
Focal Choroidal Excavation (FCE)			• **Definition:** localised area of choroidal excavation without evidence of posterior staphyloma or scleral ectasia. • **Symptoms:** Asymptomatic or mild blurring of vision or metamorphopsia. • **Fundus exam:** may be normal or show non-specific pigmentary changes. • **FAF:** irregular hypo-autofluorescence in affected area. • **OCT** shows two patterns of excavation: conforming (photoreceptor tips are in direct contact with RPE – top image) and non-conforming (photoreceptor tips are detached from RPE – bottom image). *Monitor every 6-12 months with OCT to detect any signs of macular neovascularisation. Refer to Ophthalmologist if MNV is present.*
Peripapillary Pachychoroid Syndrome (PPS)			• **Definition**: possible subset of CSCR where there is thickening of nasal choroid and peripapillary fluid pockets, typically extending from the temporal margin of the optic disc. • **Symptoms**: blur or metamorphopsia depending on location of fluid. • **Fundus exam:** peripapillary RPE mottling. Can also present with choroidal folds and ONH oedema. • **FAF**: mixed hyper and hypo-autofluoresence in affected area. • **OCT**: intraretinal and/or subretinal fluid adjacent to optic disc typically affecting nasal macula, with possible outer retinal atrophy. Can develop peripapillary CNV (seen as fibrovascular pigment epithelial detachment). *Refer to Ophthalmologist. Variable course and prognosis with no current consensus on treatment.*

CFEH TELEHEALTH
SUPPORTING OPTOMETRISTS

This reference is based on the current literature and evidence at the time of writing. This reference is designed a guide to aid diagnosis and management decisions however individual cases must be assessed in the context of all available clinical data. Management options within this reference are tailored for Australian Optometrists different scopes of practice should be considered outside of Australia. Amsler grid may be useful for self-monitoring. If you need additional advice or guidance, consider making a free telehealth appointment with a senior CFEH optometrist.

WWW.CENTREFOREYEHEALTH.COM.AU/TELEHEALTH