

Ophthalmology and Visual Sciences



Posterior Polymorphous Corneal Dystrophy (PPMD)

Category(ies): Cornea

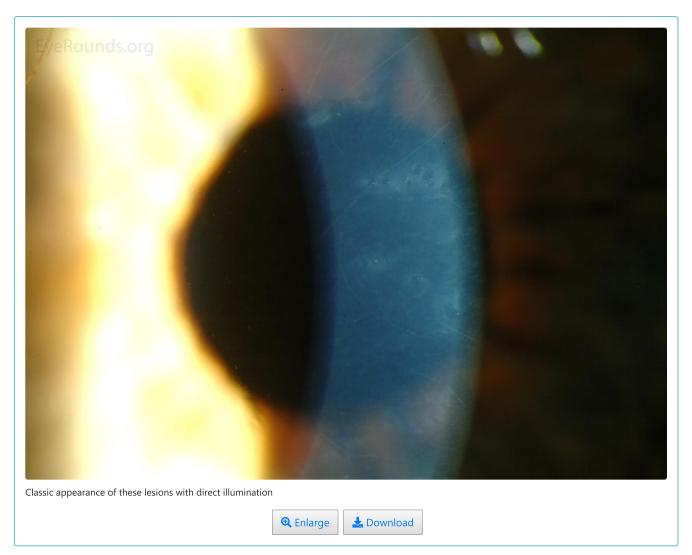
Contributor: Jesse Vislisel, MD

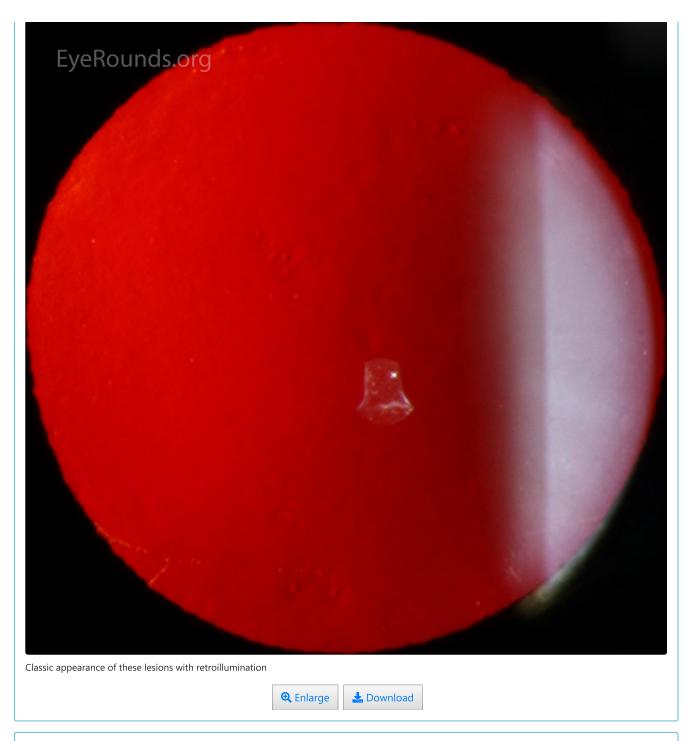
Posterior polymorphous corneal dystrophy (PPMD, PPCD) is a rare, bilateral, autosomal dominant inherited corneal dystrophy. The corneal abnormality in PPMD occurs at the level of Descemet's membrane and endothelium, and rarely will result in corneal edema or elevated intraocular pressure. The three main patterns in which PPMD may present include endothelial vesicle-like lesions, band lesions, and diffuse deep stromal opacities.

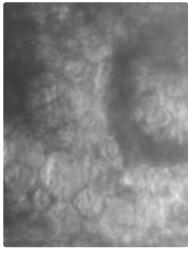
Figure 1: Vesicle-like lesions in PPMD

Contributor: <u>Jesse Vislisel, MD</u> and <u>Kenneth M. Goins, MD</u>Photographer: Brice Critser, CRA^{March 2,2015}

Vesicle-like lesions at the level of Descemet's membrane and endothelium are the hallmark lesions of PPMD. They appear as transparent cystic lesions surrounded by gray halos and commonly occur in lines or clusters. The figures below demonstrate the classic appearance of these lesions with direct illumination, retroillumination, and on specular microscopy.





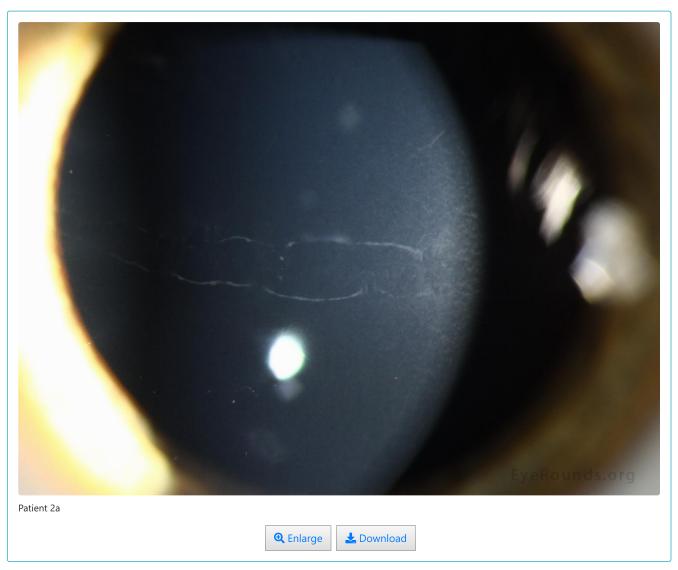


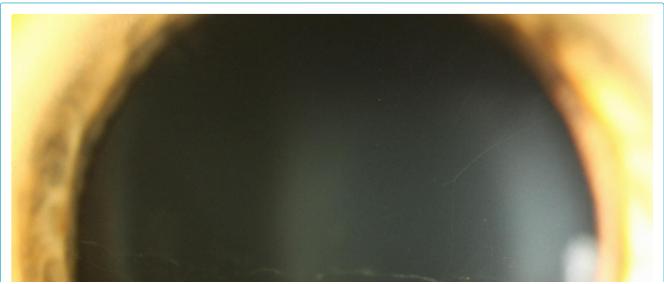
Classic appearance of these lesions with specular microscopy

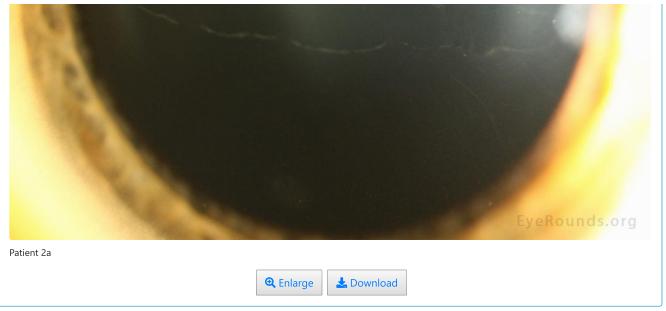
Figure 2: Band lesions in PPMD^{Contributor: <u>Jesse Vislisel, MD</u> and <u>Kenneth M. Goins, MD</u>}

Photographer: Brice Critser, CRA

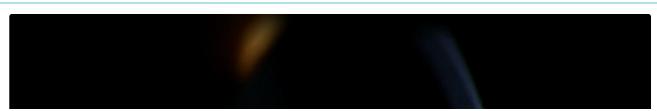
Band lesions, sometimes called "snail tracks," are classically horizontal lesions with parallel, scalloped, non-tapering edges at the level of the posterior cornea. Below are examples of these lesions in 2 separate patients.

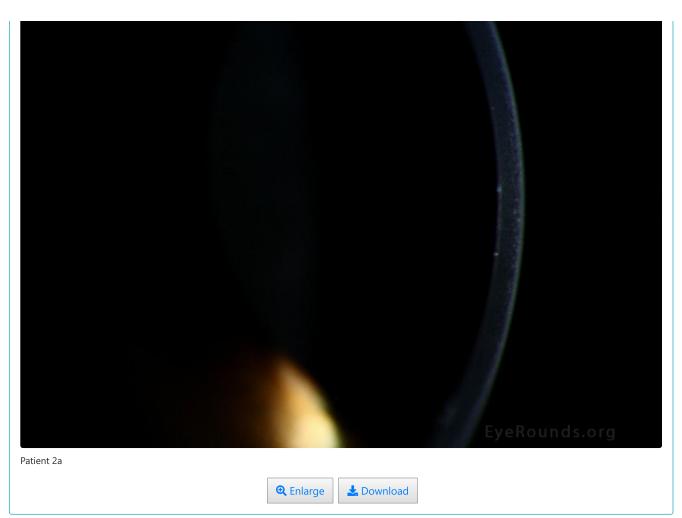


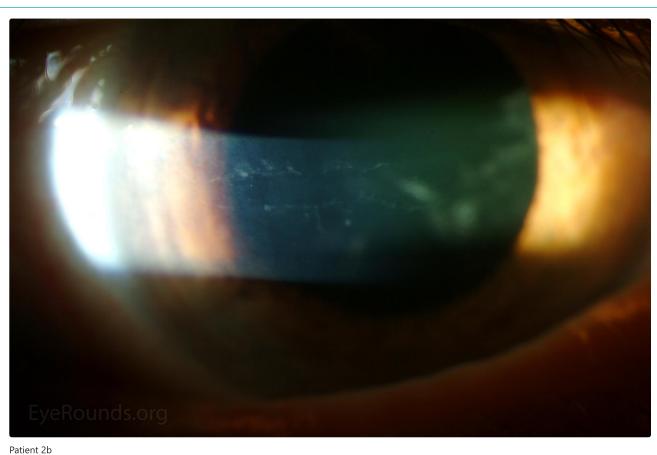












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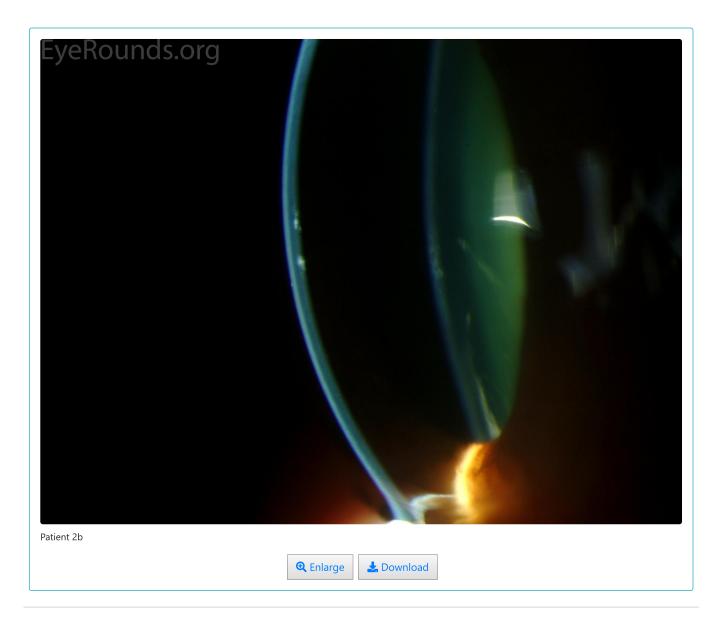
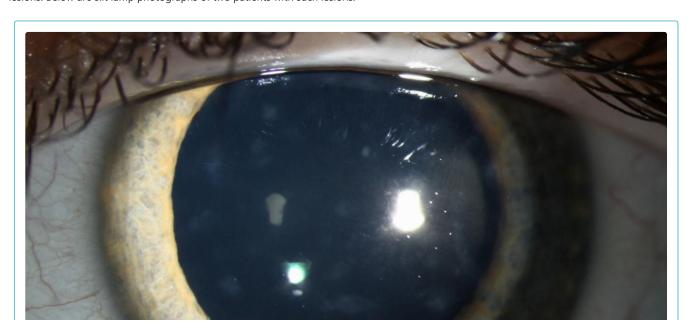


Figure 3: Diffuse posterior stromal opacities in PPMD

Contributor: Jesse Vislisel, MD and Kenneth M. Goins, MD Photographer: Brice Critser, CRA

PPMD may present with diffuse, gray-white opacities at the level of Descemet's membrane. There may be deep stromal haze adjacent to the lesions. Below are slit lamp photographs of two patients with such lesions.

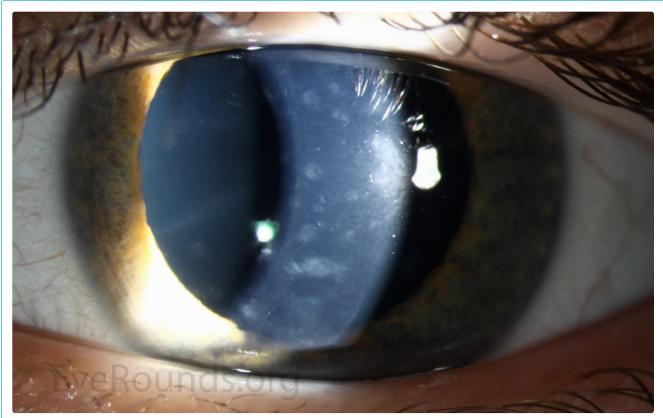




Patient 3a



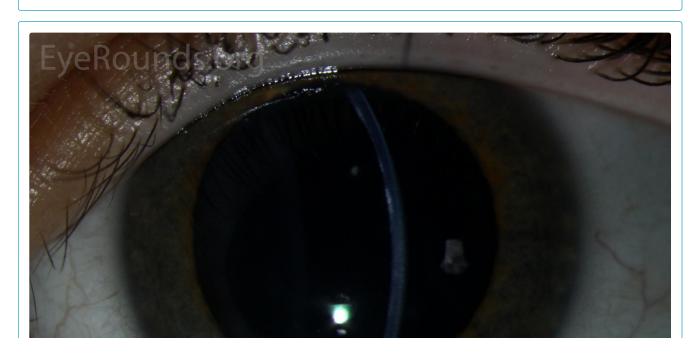


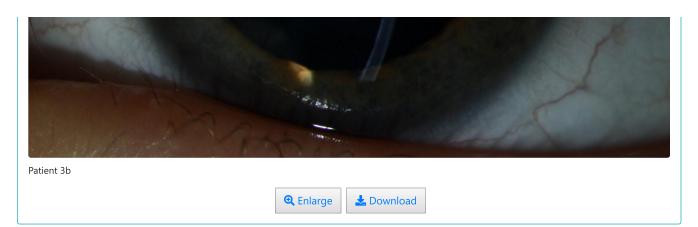


Patient 3a

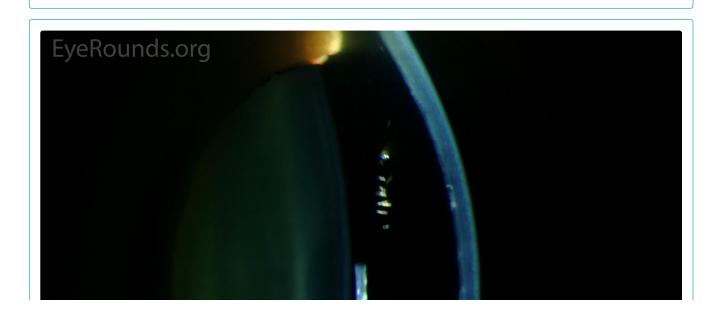












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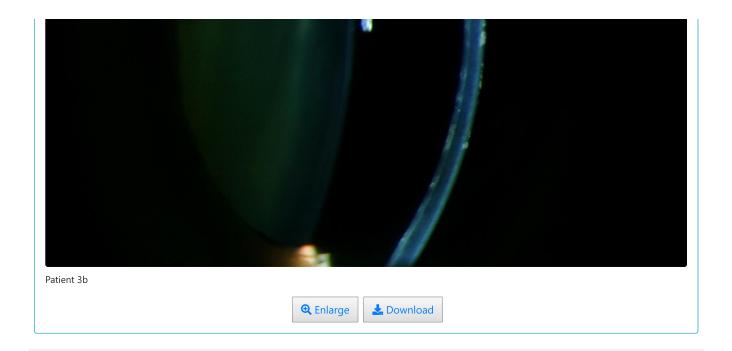


Figure 4: Confocal microscopy of affected endothelium in PPMD

Contributor: Andrew Doan, MD, PhD, University of Iowa

Confocal microscopy of the endothelium in PPMD shows epithelial-like cells. Normal endothelial cells are hexagonal and flat.

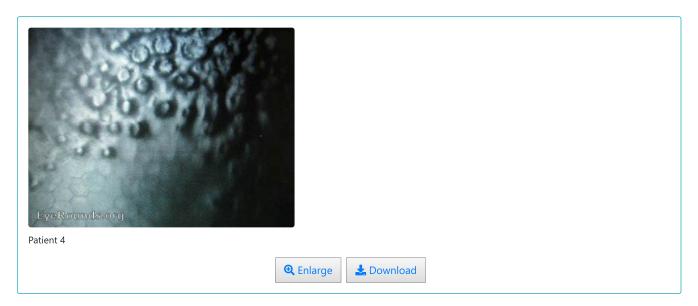


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