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.

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**Figure 1: Vesicle-like lesions in PPMD**

**Contributor:** Jesse Vislisel, MD and Kenneth M. Goins, MD

**Photographer:** Brice Critser, CRA

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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 direct illumination
Classic appearance of these lesions with retroillumination

Classic appearance of these lesions with specular microscopy
Figure 2: Band lesions in PPMD  

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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.
Figure 3: Diffuse posterior stromal opacities in PPMD

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**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 3b

Patient 3b

Patient 3b
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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