A Deeper Look: Posterior Polymorphous Dystrophy
Shalu Pal, O.D.

Posterior polymorphous dystrophy (PPD) is a slow progressing dystrophy that affects the corneal endothelium and Descemet’s membrane. PPD is a bilateral condition, though the clinical presentations are commonly quite asymmetrical.

Typical symptoms include photophobia, foreign body sensations and decreased vision. Clinical signs may be present from birth but patients may remain asymptomatic for years. The age of onset of these symptoms will determine the progression and severity of the disorder.

A slit lamp exam may reveal guttata, stromal edema and endothelial band lesions. Descemet’s membrane thickens over time diffusely or in patches. Vesicles in the cornea may form and bullous keratopathy can develop. Severe cases of PPD may result in epithelial edema and calcific and lipid degenerative changes. Iris anomalies including anterior synechiae, iridocorneal adhesions, pupillary ectropion and corectopia may also be present. IOP monitoring is important because 40 percent of patients with PPD will have elevated IOP and all those with broad-based iridocorneal adhesions will have elevated IOP.

PPD causes an abnormal layer of metaplastic endothelial cells to cover the posterior surface of the cornea, the trabecular meshwork and extend onto the iris. This layer is made of normal endothelial cells, degenerated endothelial cells, fibroblast-like cells and epithelial-like cells. These cells form irregular configurations with scalloped edges surrounded by halos giving this layer a nodular and gray geographic appearance. The irregular endothelial layer extending to the trabecular meshwork and anterior chamber angle, along with iris adhesions and anterior synechiae, are the cause of secondary open-angle glaucoma.

PPD is typically an autosomal dominant inherited dystrophy with variable expression. There are three forms of PPD that are classified based on the location of the mutated gene, associated conditions, and the severity of the condition. These are summarized in the table below:

<table>
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<th>Forms of PPD</th>
<th>PPD – 1</th>
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<td><strong>Defining characteristics</strong></td>
<td>Iris anomalies</td>
<td>Onset at birth</td>
<td>Onset varies from birth to late adulthood</td>
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<td></td>
<td>Anterior synechiae</td>
<td>Epithelial nodules and edema, Thick Descemet’s membrane</td>
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<td>Glaucoma common</td>
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<td><strong>Gene location</strong></td>
<td>VSX1 Gene</td>
<td>COL8A2 Gene</td>
<td>ZEB1 Gene</td>
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<td><strong>Associated gene mutation conditions</strong></td>
<td>Keratoconus, CHED (congenital hereditary corneal edema)</td>
<td>Early onset Fuchs’</td>
<td>Late onset Fuchs’, Inguinal hernias, Hydroceles, Bone abnormalities</td>
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<td><strong>Treatment</strong></td>
<td>Glaucoma treatment</td>
<td>PKP by age of 30</td>
<td>PKP for severe cases</td>
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To treat PPD, you need to treat the individual symptoms and complications that arise.

- Foreign body sensation
- Decreased vision
- Foreign body sensation
- Bullous keratopathy
- Corneal edema
- Glaucoma

To treat painful eyes that have otherwise good vision, consider procedures such as anterior stromal micropuncture, excimer laser phototherapeutic keratectomy, amniotic membrane transplantation, and conjunctival flap surgery.

A penetrating keratoplasty or posterior endothelial keratoplasty are reserved for patients with painful eyes and significantly reduced vision. Corneal grafts have a significant rate of failure, up to 41 percent in one study, due to regrowth of abnormal endothelial cells, graft rejection and uncontrolled glaucoma. Tip: Keep scleral lenses in mind when correcting vision post PKP!

References:


Dr. Pal received her Doctor of Optometry degree from the Southern California College of Optometry. She completed her Contact Lens and Cornea residency at the Northeastern State University Oklahoma College of Optometry where she is certified in therapeutic pharmaceutical agents, glaucoma and anterior segment lasers. Dr. Pal has a contact lens specialty practice in Toronto, Canada. She is an adjunct faculty at the University of Waterloo and a facilitator and coordinator of industry contact lens workshops at optometry schools throughout North America.

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