PIGMENTARY GLAUCOMA

Eye Learn

All about the Eye

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Pigmentary Glaucoma

1. What is “Reverse pupillary block glaucoma"? Give the clinical appearance, investigations and management of this glaucoma. D2012
2. Pigmentary glaucoma 2005

Epidemiology

- This condition constitutes 1.0–2.5% of the glaucomas seen in many Western countries.
- Pigmentary glaucoma is 3 times more common in men than in women, particularly white men who are young or middle-aged (20–50 years) and myopic**.**
- Affected women are a decade older than men.

Genetics

- Using high-resolution mapping techniques, sequencing, and functional genetic tests, Anderson showed that these conditions resulted from mutations in genes encoding melanosomal proteins.
- They postulate that pigment production and mutant melanosomal protein genes may contribute to human pigmentary glaucoma.
- Further study is needed to confirm this hypothesis.

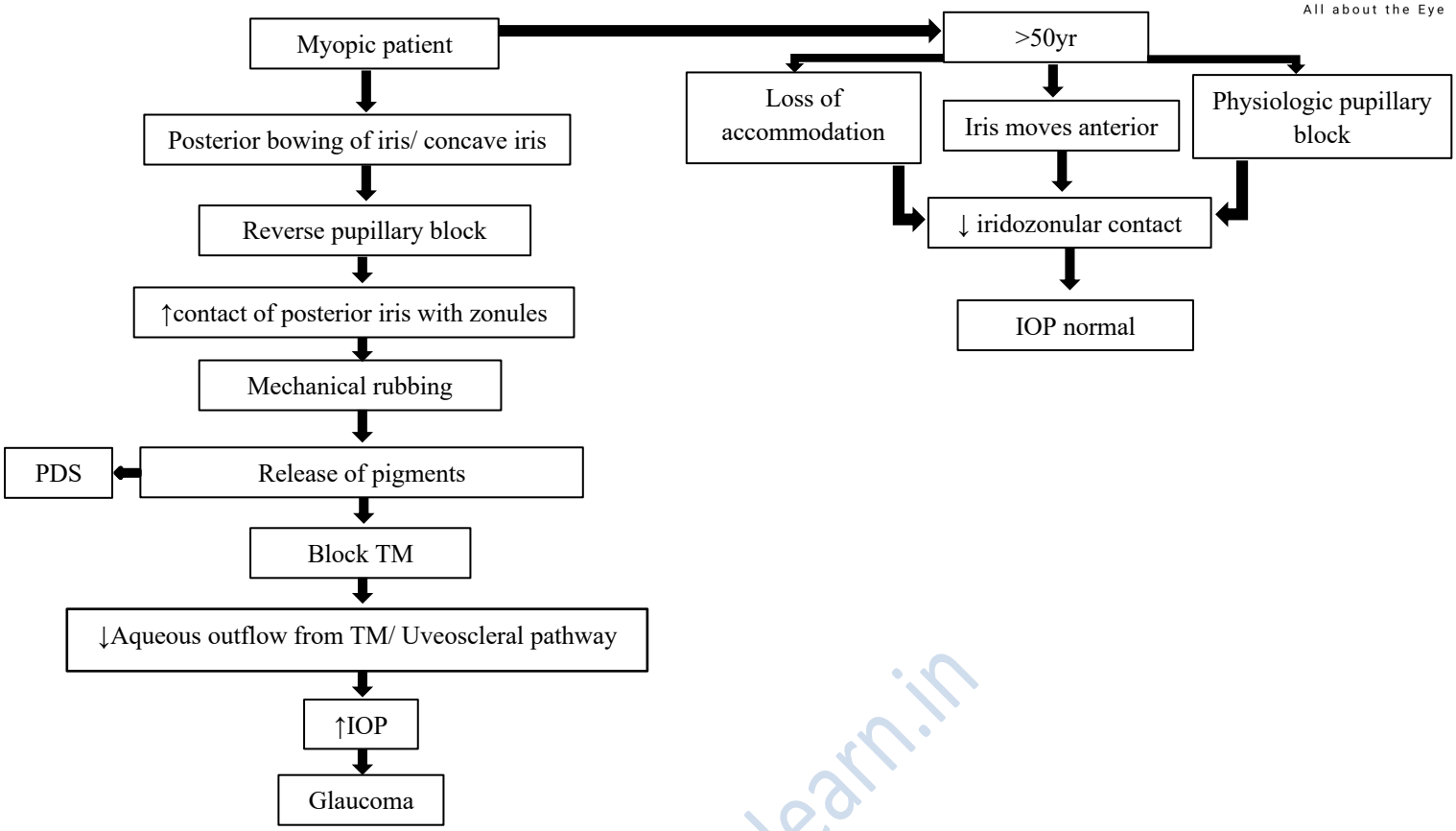
Pathogenesis

- Posterior bowing of the iris with “reverse pupillary block “configuration is noted in many eyes with pigment dispersion syndrome.
- This concave iris configuration results in greater contact with the zonular fibers, causing increased release of pigment granules.
- PDS classically presents with pigment deposits on the corneal endothelium, trabecular meshwork, anterior iris surface, zonules and lens periphery, as well as with mid-peripheral iris transillumination defects in a spoke-like pattern.
- Campbell has proposed a mechanical theory to explain pigment dispersion.
- He postulated that the concave shape of the peripheral iris allows it to rub against the zonules, causing pigment release and dispersion.
- Campbell noted that the pattern of iris transillumination defects corresponds with the arrangement of the anterior zonular packets.
- He also noted that the number and extent of the iris transillumination defects correlate with the progression of pigmentary glaucoma.
- When patients receive miotic treatment, the ensuing pupillary block lifts the peripheral iris off the zonules and allows the transillumination defects to fill in and even disappear.
- Lord and colleagues found that the pigmentary glaucoma and PDS patients had flatter keratometry than myopic controls, suggesting a difference in their anterior segment architecture.
- Not all patients with a diagnosis of pigmentary glaucoma show iridozonular contact on ultrasound biomicroscopy.
- When iris pigment is infused into animal or enucleated human eyes, outflow facility decreases and IOP increases.
- Repeated infusions of pigment, however, do not produce chronic glaucoma in animal eyes.
- Some authorities believe that patients with pigmentary glaucoma must have an underlying developmental abnormality of the outflow channels.
- As evidence for this theory, they cite
- (1) the high prevalence of prominent iris processes in patients with pigmentary glaucoma;
- (2) patients with pigmentary glaucoma who have angles that resemble infantile glaucoma, and
- (3) families who have some members with pigmentary glaucoma and other members with congenital glaucoma.

Natural history

- With increasing age, the signs of pigment dispersion may decrease as a result of normal growth of the lens, inducing a physiologic pupillary block and anterior movement of the iris.
- Loss of accommodation may also occur. As pigment dispersion is reduced, the deposited pigment may fade from the corneal endothelium, trabecular meshwork, or anterior surface of the iris.
- Approximately 15% of individuals with pigment dispersion syndrome progress to glaucoma or elevated IOP requiring treatment.
- In some cases, pigment dispersion progresses to pigmentary glaucoma overtime, which can be as long as 12–20 years.

- Remission of pigmentary dispersion also has been reported after glaucoma surgery and lens subluxation.



Histopathology

- Examination of specimens from eyes with pigmentary glaucoma demonstrates pigment and debris in the trabecular meshwork cells.
- With advanced disease, the trabecular cells degenerate and wander from their beams, allowing sclerosis and eventual fusion of the trabecular meshwork.
- Some propose that excessive phagocytosis of foreign material damages the trabecular endothelial cells and causes them to migrate.

Clinical features

1. **Iris**

- The mid-peripheral iris transillumination defects are a result of contact between the zonular fibers and the posterior iris pigment epithelium
- These defects can range in number from 1 or 2 to 65 or 70 and can be thin slits
- They are best seen in a darkened room by a dark-adapted observer.
- The pigment on the anterior iris surface accumulates in the circumferential folds and can be sufficient to give a dull or even a heterochromic appearance if the pigment dispersion is asymmetric in the two eyes.

2. **Cornea**

- The pigment deposits on the corneal endothelium are typically in a vertical spindle pattern, referred to as Krukenberg spindle, and are caused by aqueous convection currents and subsequent phagocytosis of pigment by the corneal endothelium.
- The spindle consists of extracellular as well as intracellular pigment granules phagocytized by the corneal endothelium.
- Several reports have indicated that pigment dispersion lessens with time so that Krukenberg's spindles and trabecular pigmentation become less prominent. In some cases, this is accompanied by an improvement in aqueous humor dynamics

3. **IOP**

- When the eye is dilated, pigment deposits may be seen on the zonular fibers, on the anterior hyaloid, and in the equatorial region of the lens capsule (Zentmayer ring or Scheie stripe)
- Fluctuations in IOP can exceed 50 mm Hg in untreated eyes.
- Symptoms associated with such elevated IOPs may include halos, intermittent blurry vision, and ocular pain.

Differential diagnosis

- With the exception of pigmentary dispersion, pigmentary glaucoma resembles primary open-angle glaucoma (POAG) in most aspects, including elevated intraocular pressure (IOP), decreased outflow facility, optic nerve cupping, and visual field loss.
- Large diurnal IOP fluctuations are thought to occur more often in pigmentary glaucoma and can be sufficient to cause corneal edema, blurring, and halo vision.
- Patients with pigmentary glaucoma can have a sudden release of pigment with severe IOP elevations after pupillary dilation or exercise.
- At times, this release of pigment can be confused with active anterior segment inflammation.
- Pigment release and marked IOP elevation after exercise can be blocked by topical pilocarpine therapy.
- The differential diagnosis of pigmentary glaucoma includes any condition that produces pigmentation of the trabecular meshwork.

1. normal eyes with aging,	6. previous surgery (including laser surgery),	11. herpes zoster,
2. POAG,	7. trauma,	12. megalocornea,
3. uveitis,	8. angle-closure glaucoma,	13. radiation,
4. cysts of the iris and ciliary body,	9. amyloidosis,	14. siderosis,
5. pigmented intraocular tumors	10. diabetes mellitus,	15. hemosiderosis.

- These conditions should be readily distinguished from pigmentary glaucoma by the history and physical examination.
- The condition most likely to be confused with pigmentary glaucoma is exfoliation syndrome.
- However, the pattern of iris atrophy in exfoliation syndrome is usually central and geographic, and the pigment accumulation in the trabecular meshwork consists of larger particles that are unevenly distributed about the angle.

Treatment

1. Medication

- Alpha -Adrenergic antagonists, adrenaline (epinephrine), dipivefrin, and carbonic anhydrase inhibitors (CAIs) are useful in the management of pigmentary glaucoma.
- Miotic agents reduce IOP in pigmentary glaucoma and are theoretically appealing because they increase pupillary block and lift the peripheral iris from the zonules.
- Reports also indicate that patients with pigmentary glaucoma have a high incidence of retinal detachment; thus a careful peripheral retinal examination is mandatory before cholinergic agents can be prescribed.

2. **PI**

- Laser peripheral iridotomy has been proposed as a means of minimizing posterior bowing of the iris peripheral iridectomy to cure the 'reverse' pupillary block that is responsible for the characteristic peripheral iris concavity.
- Ultrasonic biomicroscopy is helpful in indicating those eyes that are most likely to beneft from iridectomy.
- Perhaps iridectomy may help during phases of the syndrome when pigment granules are being liberated actively, but iridectomy seems unlikely to benefit established cases when the damage has been done.
- Ideally peripheral iridectomy would provide effective prophylaxis for patients with PDS before they develop glaucomatous visual field loss.

3. **ALT**

- Should be performed. Because of the heavy pigmentation of the angle, ALT is done with relatively low energy settings in the range of 200–600mW. Selective laser trabeculoplasty also works.

4. **Filtering surgery** is usually successful; however, extra care is warranted in young myopic male patients, who are at increased risk for hypotony maculopathy

- Despite the young age of these patients, the results of surgery are generally successful.