Ultrasound Biomicroscopy in Asymmetric Pigment Dispersion Syndrome and Pigmentary Glaucoma

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Objective: To identify differences in anterior chamber anatomy among patients with asymmetric pigment dispersion syndrome and no other discernible cause for the asymmetry.

Methods: Ultrasound biomicroscopy and A-scan biometry were performed on both eyes of 13 patients with asymmetric pigment dispersion syndrome without a known cause for asymmetric involvement. A radial perpendicular image in the horizontal temporal meridian detailing the scleral spur, angle anatomy, and iris configuration was obtained for each eye by 2 examiners.

Results: There were no differences in lens thickness (P=.33), refractive error (P=.84), or axial length (P=.99) between more and less affected eyes. However, the mean ± SD iris concavity (P<.001), iris-lens contact distance (P=.02), and distance from the scleral spur to the iris insertion (0.42±0.11 vs 0.29±0.06 mm) (P=.002) were greater in the more affected eye of each patient.

Conclusion: A more posterior iris insertion predisposes to the phenotypic expression of pigment dispersion syndrome.

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In pigment dispersion syndrome (PDS), friction between the posterior iris surface and the anterior zonular bundles causes the disintegration of iris pigment epithelial cells and the release of pigment granules, which are then dispersed by aqueous currents. The liberated pigment is deposited throughout the anterior segment. The classic diagnostic triad consists of a Krukenberg spindle, slitlike radial midperipheral iris transillumination defects, and increased pigmentation of the trabecular meshwork. The angle is typically wide open, the iris is inserted posteriorly, and the configuration of the peripheral iris is posteriorly concave. Elevated intraocular pressure develops in many patients and may lead to glaucomatous damage (pigmentary glaucoma).

Pigment dispersion syndrome is an autosomal dominant disorder. Myopia predisposes to its phenotypic expression and is present in approximately 80% of affected patients. The most significant risk factors for the development of the phenotypic expression of PDS are young age, male sex, myopia, white race/ethnicity, and a positive family history. Although men and women are equally affected, men are more likely to develop glaucoma, in approximately a 3:1 ratio.

Phenotypic expression is typically bilateral and symmetric. A marked asymmetric involvement is unusual, and a cause should be sought when present. Asymmetry may occur because a second condition, such as cataract formation or extraction, Horner syndrome, or Adie pupil limits the involvement in 1 eye or because the occurrence or development of exfoliation syndrome or angle recession led to higher intraocular pressure and more frequent glaucoma in the doubly involved eye. In some patients, no reason for asymmetry can be discovered even after a thorough clinical examination. In this article, we describe 13 such patients in whom a more posterior iris insertion in the more involved eye was detected by ultrasound biomicroscopy (UBM).

METHODS

High-frequency high-resolution anterior segment UBM (UBM P-40; Paradigm Medical Industries, Salt Lake City, Utah) and A-scan biometry (A/B scan; Sonomed Inc, Lake Success, NY) were performed before pharmacologic pupillary manipulation in both eyes of all patients with asymmetric PDS without discernible cause of the asymmetry. A sagittal image in the horizontal temporal meridian in both
eyes detailing the Schwalbe line, scleral spur, and iris root inser-
tion was obtained for each eye by 2 masked examiners (S.D. and C.T.) under standardized room lighting conditions. Measure-
ments were made using the software UBM Pro2000 Paradigm Medical Industries). Corneal diameter was measured using a handheld caliper.

The following 3 landmarks were used as reference points for UBM measurements (Figure 1 and Figure 2): (1) the Schwalbe line (the termination of Descemet membrane, which appears as a hyperreflective line in the posterior aspect of the cornea), (2) the scleral spur (a hyperlucent wedge at the an-
terior edge of a line separating corneal tissue and ciliary muscle fibers [Figure 1]), and (3) the iris root insertion (the anterior-most insertion of the iris into the ciliary body).

The UBM image measurements were made by a masked ob-
server (S.D. or C.T.) using a random image order. Variables mea-
sured included the linear extent of iris-lens contact distance (ILCD), iris configuration (Figure 2 and Figure 3), and distance from the scleral spur to the iris insertion. The iris configuration was measured by drawing a line connecting the most peripheral and most central points of the iris pigment epithelium (reference line) and by measuring the largest perpendicular distance from the line to the iris pigment epithelium. A concave or convex surface was determined to exist when there was a measurable difference between the plane of the iris pigment epithelium and the reference line. Negative values were assigned to concave irides, and positive values were assigned to convex irides. A planar iris received a 0 value. Measurements of lens thickness and axial length were obtained using A-scan biometry.

Statistical evaluation was performed using t test when evaluat-
ing differences between more and less affected eyes. The soft-
ware program JMP 4.0 was used (SAS Institute Inc, Cary, NC).

RESULTS

Eleven men and 2 women were included in the study. The mean ± SD patient age was 44.9±7.3 years. Only 1 patient did not have higher intraocular pressure in the more affected eye, and 9 of 13 patients had glaucoma-tous optic neuropathy in at least 1 eye.

Patients’ intraocular pressure and cup-disc ratio are given in Table 1. All patients had 20/20 visual acuity. The mean ± SD horizontal corneal diameters were 12.7±0.3 and 12.6±0.5 mm, lens thicknesses were 4.24±0.45 and 4.01±0.39 mm, and axial lengths were 24.7±0.7 and 24.7±0.8 mm in more and less affected eyes, respectively (P > .05 for all).

The UBM measurements are given in Table 2. There was a more concave iris configuration (−0.28 vs 0.08 mm), increased iridolenticular contact (1.44 vs 0.91 mm), and greater distance from the scleral spur to the iris insertion (0.42 vs 0.29 mm) in the eyes with more severe PDS. There was no difference in lens thickness or axial length to account for the asymmetry of the anterior segment appearance.
We performed Pearson product moment correlation, looking for any relationship between age, lens thickness, ILCD, and anterior chamber depth. We found a positive correlation between age and lens thickness in affected ($r = 0.29$) and unaffected ($r = 0.21$) eyes and found a comparatively more negative correlation between ILCD and anterior chamber depth in affected ($r = -0.68$) eyes than in unaffected ($r = -0.19$) eyes. However, there was no significant relationship between lens thickness and ILCD ($r = -0.01$).

**COMMENT**

The proximate causal abnormality responsible for the pathogenesis of PDS is iridozonular friction, thought to result from backward bowing of the peripheral iris, bringing it into contact with the anterior zonular bundles. A similar concavity can be induced by accommodation in young healthy individuals and in individuals with PDS. Therefore, the degree of iris concavity seems to be a dynamic state.

The cause of the iris concavity in PDS remains unknown. Campbell and Karickhoff noted that laser iridotomy eliminates the concavity and hypothesized that a reverse pupillary block mechanism exists in which the iris drapes over the lens and acts as a flap valve that prevents aqueous in the anterior chamber from returning to the posterior chamber. This has been confirmed by UBM. The pressure in the anterior chamber then exceeds that of the posterior chamber, pushing the iris posteriorly, creating a concave configuration, and bringing the iris pigment epithelium into contact with the zonular bundles. The greater the contact, the greater should be the pigment dispersion. Patients with asymmetric PDS offer a unique opportunity to evaluate anatomic differences between more and less affected eyes.

**Table 1. Patients’ General Features**

<table>
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<th>Patient No.</th>
<th>Age, y</th>
<th>Intraocular Pressure, mm Hg</th>
<th>Cup-Disc Ratio</th>
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<td>More Affected Eye</td>
<td>Less Affected Eye</td>
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**Table 2. Ultrasound Biomicroscopy Measurements**

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<tr>
<td>Scleral Spur–Iris Root Distance*</td>
<td>0.42</td>
<td>0.29</td>
<td>-0.28</td>
<td>0.08</td>
<td>1.44</td>
<td>0.91</td>
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<td>Iris Configuration†</td>
<td>0.11</td>
<td>0.06</td>
<td>0.15</td>
<td>0.25</td>
<td>0.43</td>
<td>0.57</td>
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<td>Iris-Lens Contact Distance‡</td>
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*P = .002.
†P < .001.
‡P = .02.
ously shown that the insertion of the iris into the ciliary body is more posterior in eyes with PDS than in control eyes.2,24,26-28

In the present series, the more affected eye of our patients had a more posterior insertion of the iris root. The statistical significance attained (P = .05) between more and less affected eyes is striking given the small sample size. We suggest that the more posteriorly inserted iris is in greater contact with the zonular apparatus, leading in turn to more prominent manifestations of PDS. Sokol et al20 found a significant difference in the mean ± SD distance from the scleral spur to the iris insertion between patients with PDS (0.40 ± 0.04 mm) and control subjects (0.28 ± 0.04 mm); however, they did not measure the iris configuration. In our study, the mean ± SD distance from the scleral spur to the iris insertion was statistically greater in the more affected eyes (0.42 ± 0.11 mm) compared with the less affected eyes (0.29 ± 0.06 mm). We also found a greater iris concavity in the more affected eyes. The presence of increased negative correlation of ILCD with anterior chamber depth in affected eyes compared with unaffected eyes confirms a larger pressure gradient between the anterior chamber and the posterior chamber in affected eyes compared with unaffected eyes. In unaffected eyes, the reverse pupillary block mechanism is incomplete, and there is less pressure differential and less ILCD. This also confirms the reverse pupillary block mechanism for the development of PDS described by Campbell.2

Pigment dispersion syndrome has been associated with large corneas.29,30 However, the measurement of corneal Campbell.2 mechanism for the development of PDS described by ILCD. This also confirms the reverse pupillary block complete, and there is less pressure differential and less sex differences in pigmentary glaucoma. Bick MW. Sex differences in pigmentary glaucoma. Am J Ophthalmol. 1986;54:831-837.


