Imaging Zonular Abnormalities Using Ultrasound Biomicroscopy

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Objectives: To evaluate the use of ultrasound biomicroscopy (UBM) in detecting ciliary zonular defects and to describe the UBM signs of such defects.

Methods: Eighteen eyes of 18 patients with clinically suspected zonular abnormalities were evaluated using UBM. Predisposing factors included pseudoexfoliation, congenital spherophakia, surgical procedure, trauma, and the Marfan syndrome.

Results: Of 18 eyes, 11 showed UBM evidence of missing zonules and 11 showed evidence of zonular stretch. Four of the 18 eyes had both missing and stretched zonular fibers. All of the eyes examined showed increased lenticular sphericity in the area of zonular disorder. Nine eyes showed ciliary body flattening. Pupillary block was seen in 5 patients, and angle crowding due to direct iridal rotation was noted in 3.

Conclusions: Ultrasound biomicroscopy can detect zonular loss and stretching directly. Increased lenticular sphericity and ciliary body flattening are signs of zonular defects. Angle closure mechanisms include pupillary block and direct iridal rotation.

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PATIENTS, MATERIALS, AND METHODS

We performed a retrospective review of 18 eyes of 18 patients with clinically suspected zonular abnormalities who were examined using UBM. The patients’ ages ranged from 12 to 80 years; 8 were male and 10 were female. Clinical factors that led clinicians to recognize zonular abnormalities included signs of increased lenticular mobility, direct visualization of part of a disrupted zonule, or a history of a condition in which zonular abnormalities are frequently present.

The equipment used was a commercial version of the original equipment described by Pavlin and colleagues as developed by Humphrey Instruments (San Leandro, Calif). A 50-MHz transducer was used, which allowed a penetration depth of approximately 4 mm and a resolution of 40 µm. After informed consent was obtained, all scanning was performed with the patient in the supine position under the same room-lighting conditions. Radial images were taken through the lens, zonular bundles, and the ciliary body. A normal anterior zonule appears as a medium reflective line extending from the ciliary process to the lens margin (Figure 1). The anterior zonular fibers are always imaged when appropriate techniques are used. These techniques include placing the focal zone of the transducer at the anterior zonular region, orienting the scanning plane perpendicular to the zonular path, and moving the probe slightly to optimize sound reflection from this structure. The posterior zonular fibers frequently cannot be detected because of difficulty scanning in this region due to interference from the lens margin.

was responsible in 5 of these 8 eyes (Figure 7). In the remaining 3 eyes, the angle narrowing was produced by direct anterior iridal rotation in the absence of pupillary block (Figure 8).

Figure 1. An ultrasound biomicroscopic image of a normal angle showing the relationship of the zonule to the lens and ciliary body. The zonule (arrow) extends to the lens surface, which has a relatively flat profile in this region.

Figure 2. An ultrasound biomicroscopic image of the eye of a patient with the Marfan syndrome showing an area of missing zonules (arrow) and increased distance between the lens and ciliary body.

Figure 3. An ultrasound biomicroscopic image of the eye of a patient with spherophakia showing zonular stretch (arrow).

Figure 4. An ultrasound biomicroscopic image of the eye of a patient with pseudoexfoliation showing zonular remnants on the lens margin (arrow).

The ciliary body and zonular fibers have previously been imaged with UBM. This study confirms the ability...
of UBM to consistently and accurately identify and localize zonular abnormalities. This direct approach can be valuable in confirming clinical impressions based on indirect signs. The ability to determine the degree and extent of zonular abnormalities could be of value in planning the surgical approach to cataract removal. In addition, UBM helps define the mechanism of angle closure induced by zonular weakness and changes in lenticular configuration and position. Subtle weakness in the zonular structure, however, is unlikely to be detected by UBM.

The patients in our study had diverse causes of zonular abnormalities. Ultrasound biomicroscopy was consistently able to accurately identify and localize these defects. The UBM signs of zonular abnormality can be divided into direct and indirect signs. The direct signs of zonular defects include missing zonular fibers, increased zonular fiber length, and the presence of zonular remnants on the lens capsule. The indirect signs include increased lenticular sphericity, ciliary body flattening, and increased lens–ciliary body distance. The increase in regional lenticular sphericity is of interest. The loss of zonular tension produced a tendency for the lens to assume a more spherical form, a process that is likely an exaggeration of the normal lenticular changes that occur with accommodation.

In our study, trauma was the most commonly identified cause of zonular defects. Nishikawa and Okisaka\textsuperscript{6} have shown that younger patients have stronger zonules than older patients. In addition, when excessive force is applied, the zonules tend to break in the region adjacent to the ciliary body in younger patients, whereas in older patients, the break occurs adjacent to the lenticular margin. Our patients with trauma showed areas of missing zonules, but the point of rupture could not be determined because zonular remnants could not be identified. Three patients with trauma showed localized areas of absent zonular fibers. Of interest is that in the area of missing zonular fibers, the lens appeared more curved than the area with intact zonules. The transition zone between intact and missing zonular fibers could be clearly defined and designated in clock hours. These features were also noted in the 2 patients who had had iridocyclectomy.

Three of our patients had the Marfan syndrome. The embryonic development of the zonule takes place during the third and fourth months.\textsuperscript{7} Genetic alterations in
conditions such as the Marfan syndrome can result in zonular abnormalities. Lenticular dislocation occurs in approximately 80% of patients with the Marfan syndrome.8-10 Two of our patients with the syndrome showed large areas of missing zonules with lenticular dislocation. The ciliary body was flattened in the region of missing zonules.

The pseudoexfoliation syndrome is associated with a high incidence of phacodonesis, spontaneous lenticu-
tion. The ciliary body was flattened in the region of miss-
large areas of missing zonules with lenticular disloca-
tion. It is of interest that our patient with pseudoexfoliation showed prominent zonular remnants attached to the lens capsule. This may imply that the source of rupture is more peripheral in these patients, either at the ciliary process or midzonule.

The zonules in spheroophakia have been described as being elongated.11 The UBM signs of spheroophakia have been described previously.11,12 We describe 3 more pa-
tients in our study, all of whom showed stretched zon-
ules. The ciliary processes in the patients with sphero-
phakia appeared of normal length but thinner and less well formed than normal. The steep anterior lenticular curvature and stretched zonular fibers were easily noted on UBM in all of our patients. All 3 patients showed narrow angles, 1 due to pupillary block and 2 due to direct iridal rotation.

Anterior chamber angle narrowing or closure may be causally or coincidentally associated with zonular de-
fects. Ultrasound biomicroscopy is useful in establish-
ing a diagnosis of a narrow or closed angle and can help define the mechanism of angle narrowing. Pupillary block is produced when the iris-lens interface is anterior to the iridal root. This can occur with the increased lenticular sphericity that occurs with extensive zonular loss. The UBM hallmark of this mechanism is an anterior bowing of the iris caused by the pressure differential between the posterior and anterior chambers. Three of our patients, however, displayed angle narrowing in the absence of iri-
dal convexity. In these patients, 2 with congenital sphero-
phakia with extremely shallow anterior chambers and 1 with trauma, the angle was narrowed by direct ante-
rior rotation of the iris. The iris in these patients was plan-
lar, with the tip of the iris being supported by the ante-
riorly displaced lenticular surface and the iris rotated forward at its root, producing contact between the ante-
rior iridal surface and the trabecular meshwork. This mechanism of angle closure has not previously been clearly defined. The importance of differentiating this mechanism from pupillary block relates to the likely-
hood that these patients will not respond to iridotomy.

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REFERENCES

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uly 13, 1895, the operation was decided upon and performed by Prof. Weiss and Dr. Heuck in the hospital at Mann-
heim. The child having been chloroformed, a cutaneous incision was made diagonally from above outwards, down-
wards, and inwards in such a way that the height of its convexity reached the middle of the outer edge of the orbit. The incision was carried down to the fascia and periosteum, which were divided close to the edge of the orbit. . . . After dividing the periosteum the external angular process of the frontal bone and the frontal process of the malar were chiseled through toward the infraorbital fissure. . . . Following this, the periorbital tissues were opened longitudinally. A tense, tendinous sack of bluish lustre at once appeared in the wound, and being seized with forceps was readily separated in its anterior por-
tion.

These findings, together with the exophthalmus, indicated plainly a tumor deep in the orbit. All evidence of an inflam-
atory process, fever, pain, redness and swelling, was absence of pulsation and bruit. Taking the age of the child into con-
sideration, the first thought would be of a tumor of the optic nerve.