IMAGES IN CLINICAL MEDICINE

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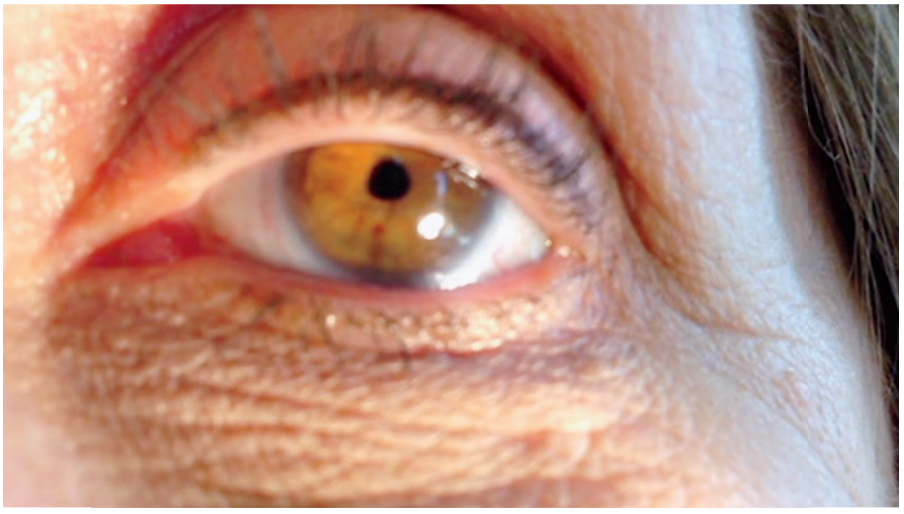
Iridodonesis



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A video showing iridodonesis is available at NEJM.org

A 65-year-old woman presented to a clinic to be evaluated for Marfan's syndrome after her identical twin received a diagnosis of the disorder. Physical examination revealed iridodonesis, or "dancing" of the iris, which was elicited by rapid movement of the eye (see video). A 3/6 holodiastolic murmur was also noted. Echocardiography revealed dilatation of the sinus of Valsalva (to a diameter of 5.0 cm), severe aortic regurgitation, and mitral-valve prolapse. Genetic testing revealed a mutation (Cys576Gly variant) in the gene encoding fibrillin, which confirmed the diagnosis of Marfan's syndrome. Iridodonesis, which is described as a shimmering or tremulousness of the iris that occurs with eye movement, is a sign of ectopia lentis, the total or partial dislocation of the lens that is a characteristic sign in patients with Marfan's syndrome. Fibrillin, a major component of the zonules, is responsible for stabilizing the lens within the eye; mutation of the gene encoding fibrillin can weaken the zonules and lead to ectopia lentis. The patient underwent successful replacement of the aortic valve and root and was doing well at her 3-year follow-up visit. She also has regular follow-up visits with her ophthalmologist, with no further treatment undertaken.

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