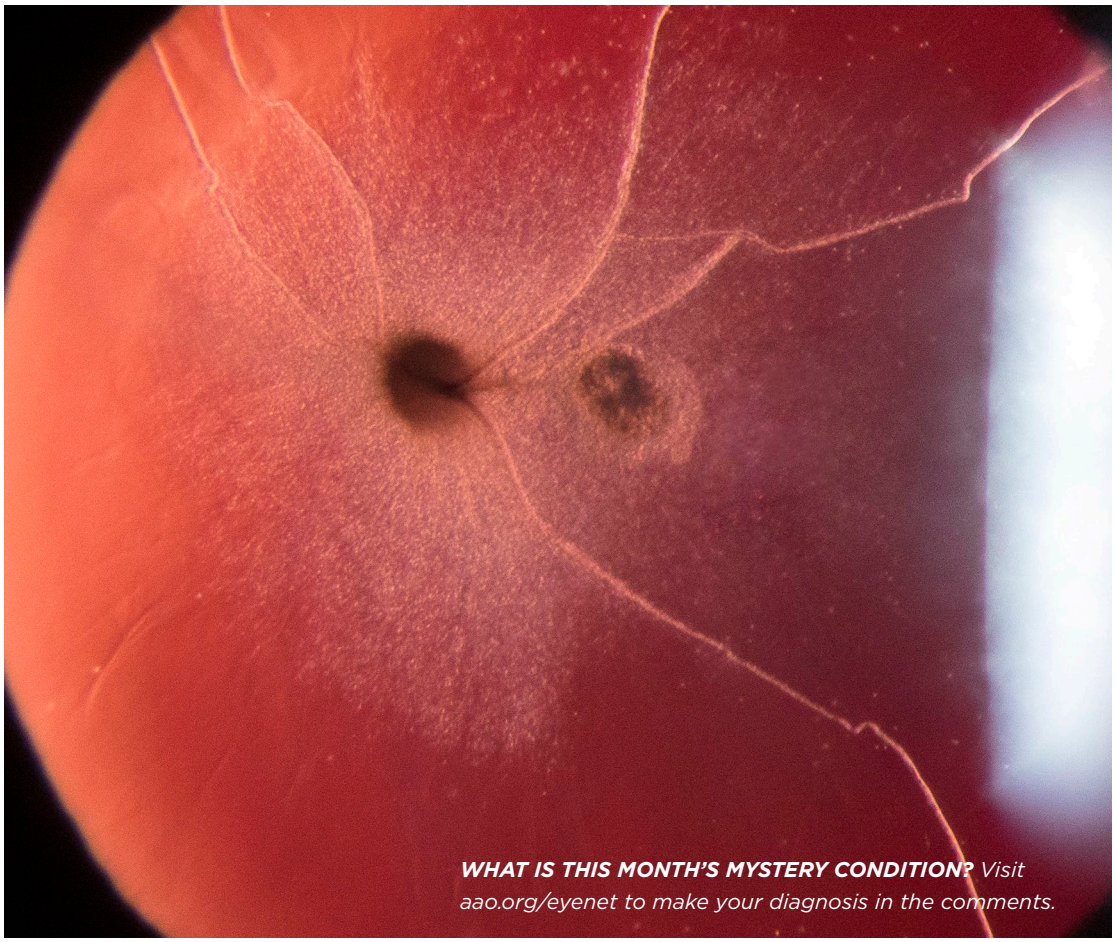
MYSTERY IMAGE

BLINK



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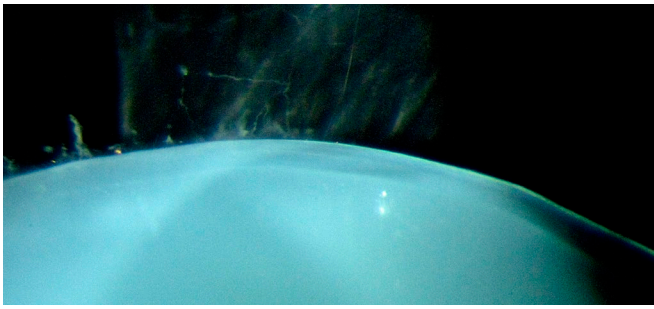
LAST MONTH'S BLINK

Ectopia Lentis in a Patient With Homocystinuria

A 9-year-old girl had been diagnosed with homocystinuria as an infant and, over the years, had ophthalmic screening for ocular associations of the disorder. She was found to have bilateral inferior subluxation of clear lenses. By age 9, the subluxation had progressively worsened (see photo) such that it was affecting her aided vision significantly. Her visual acuity was 20/63 in the right eye and 20/50 in the left.

Homocystinuria is an autosomal recessive inherited disorder of methionine metabolism due to deficiency of cystathionine beta-synthase. The zonule normally contains high levels of cystine, and a deficiency of this amino acid leads to increased fragility of the zonular fibers, which then alters the lens stability.

Of interest in this photograph of the right eye are the curly ends of the broken zonular fibers seen at the lens equator of the subluxated lens.



This is an important differentiating feature for ectopia lentis seen in Marfan syndrome. In that setting, these fibers are abnormally elongated but not fragile and broken.

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