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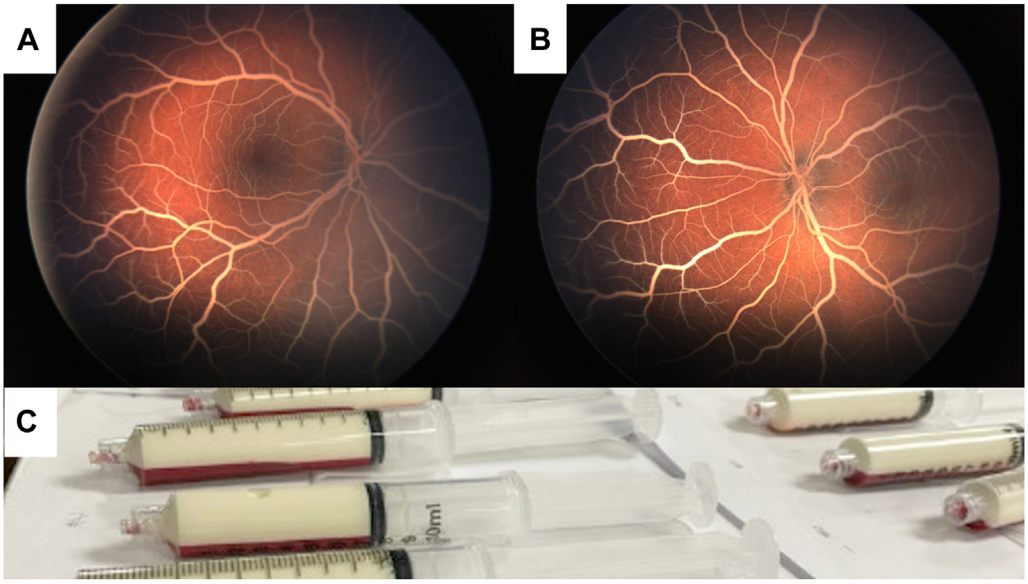
Pictures & Perspectives



Severe Lipemia Retinalis in an Infant with Autosomal Recessive Lipoprotein Lipase Deficiency

A 40-day-old female infant presented with 2 weeks of poor feeding, melena, and rectal bleeding. Examination revealed bilateral severe lipemia retinalis, characterized by bright white retinal vessels over a "salmon-colored" fundus (**A**, **B**). Triglyceride levels were extremely elevated (437.05 mmol, reference range $\leq$ 1.12 mmol). Genetic testing confirmed an autosomal recessive mutation causing lipoprotein lipase deficiency (**C**). The retinal appearance is the result of extremely high circulating serum chylomicrons levels. This case highlights a rare cause of genetic hyperlipidemia with dramatic fundus manifestations but typically without afferent visual dysfunction. (Magnified version of Figure **A**-**C** is available online at www.aaojournal.org).

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