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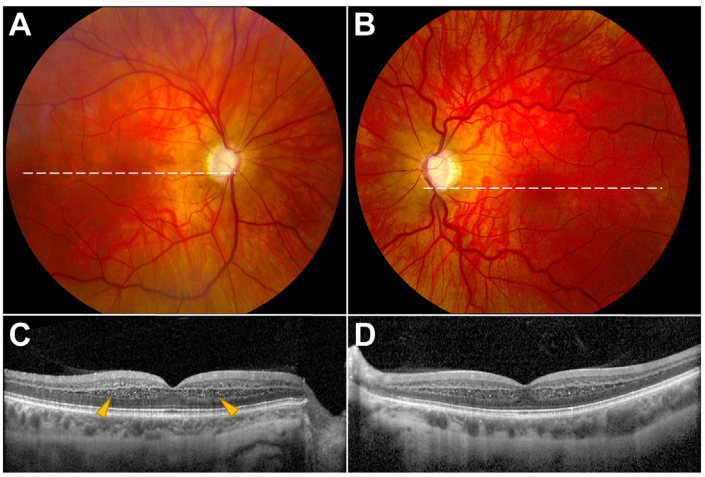
Pictures & Perspectives



Peculiar Outer Plexiform Layer in Autosomal Dominant Wolfram Syndrome

A 29-year-old man with congenital sensorineural hearing loss and type 1 diabetes was referred for evaluation of macular edema. Examination revealed pallorous, excavated optic nerve heads with blunted foveal reflex in both eyes (**A**, **B**). OCT demonstrated multilaminated outer plexiform layer (OPL) with hyperreflective foci (**C**, **D**; arrowheads). Genetic testing revealed a de novo heterozygous missense mutation in *WFS1* (Ala684Val), consistent with Wolfram syndrome. This peculiar OPL clefting is only seen in dominant *WFS1* missense mutations and is thought to result from deranged Muller cell architecture and contracted synaptic pedicles in the OPL because of abnormal wolframin expression. (Magnified version of Figure **A**-**D** is available online at www.ophthalmologyretina.org).

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