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Ophthalmology and Visual Sciences

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Batten Disease

Category(ies): Genetics, Inherited Eye Disease, Retina

Contributor: [Jeffrey D. Welder, MD](#)

Photographer: Carol Chan, CRA

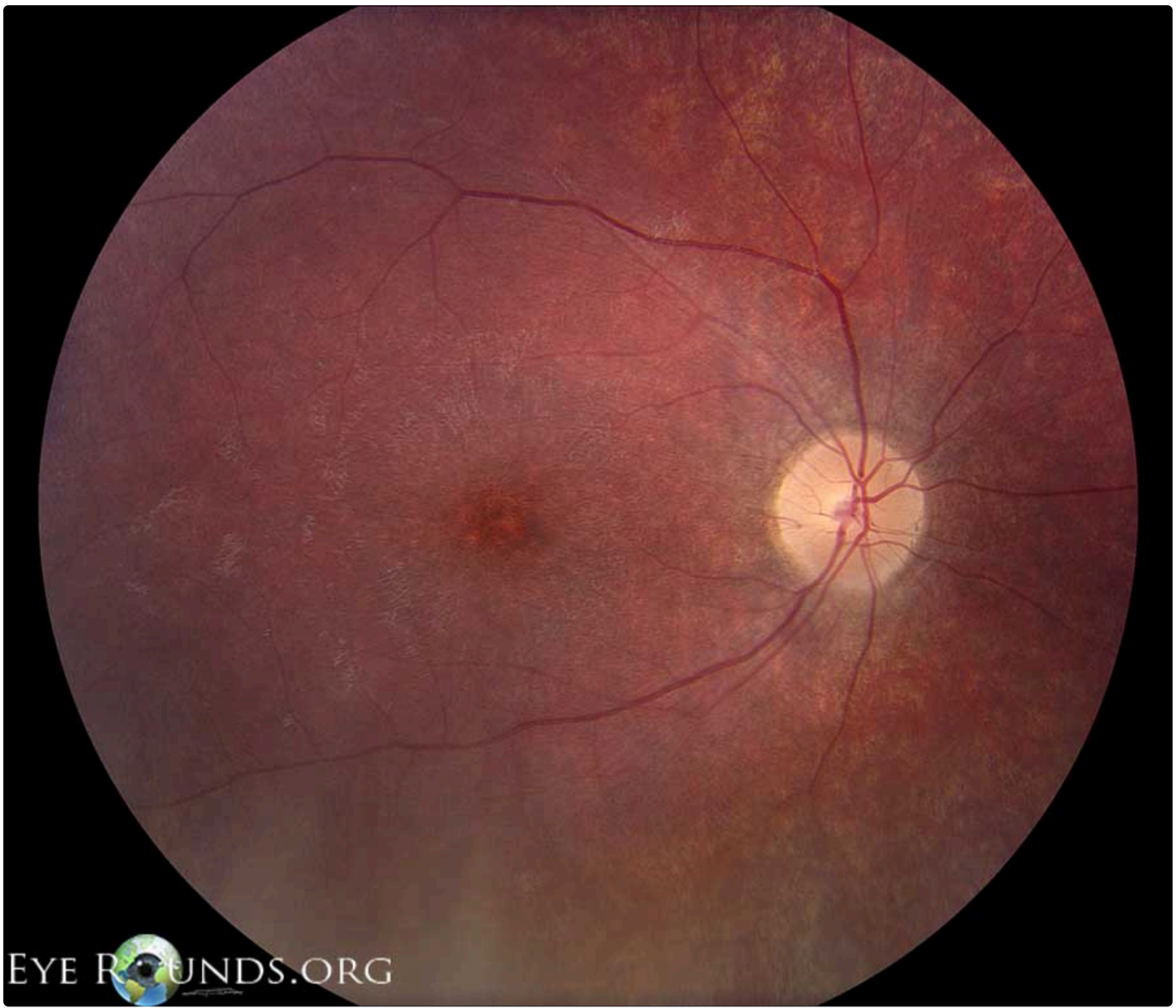
Posted: June 19, 2013



7-year-old female with molecularly confirmed CLN3-associated Batten Disease.

Fundus photos demonstrate mild optic nerve pallor, central macular mottling, attenuated arterioles and peripheral retinal granularity. There was 1+ vitreous cell present. Vision was 20/800 in the right eye and hand motion in the left. Batten disease is a fatal autosomal recessive neurodegenerative disease and is the most common neuronal ceroid lipofuscinosis.

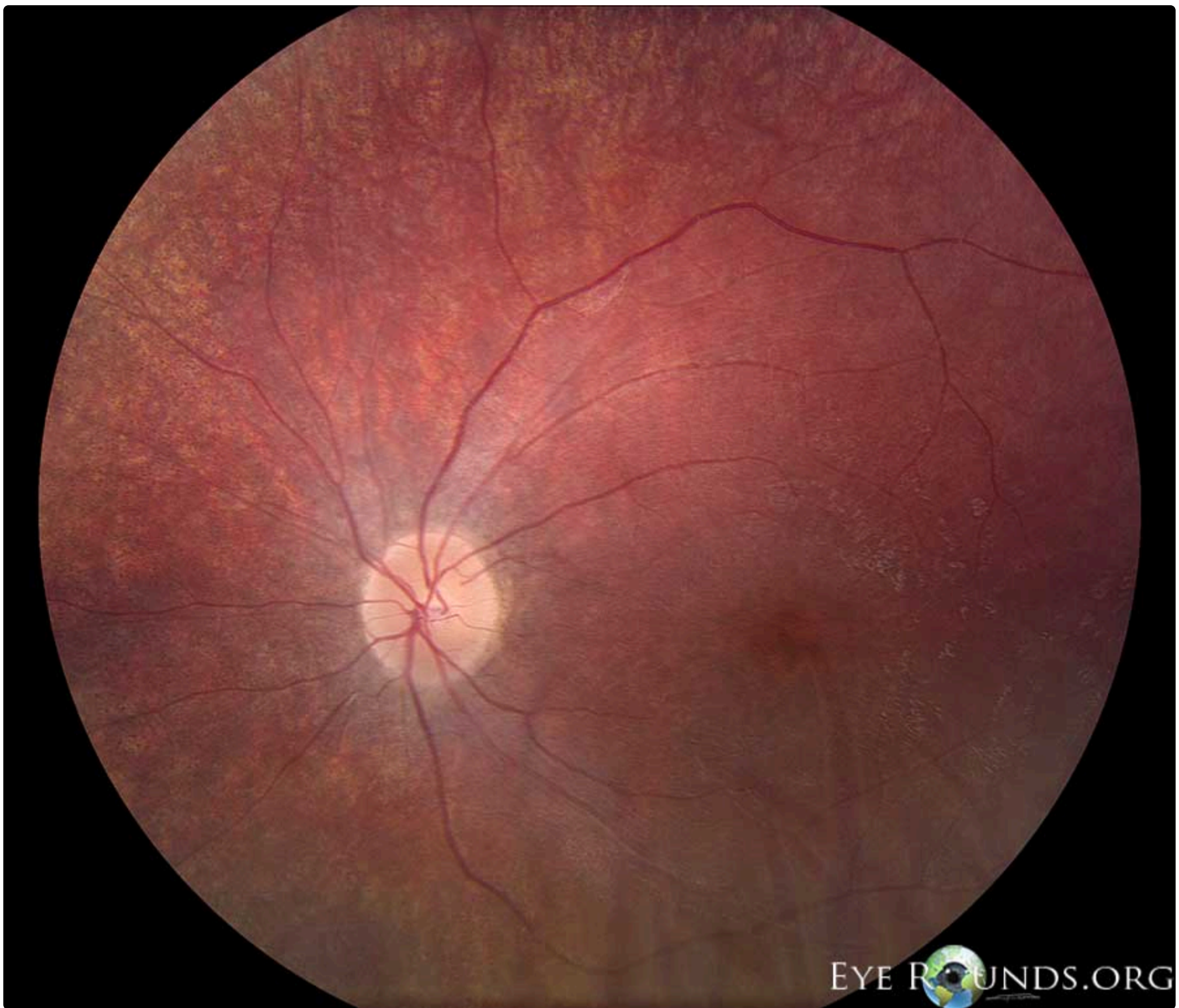
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Right eye

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Left eye

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Bullseye maculopathy in Batten Disease

Category(ies): Inherited Eye Diseases, Retina / Vitreous

Contributor: Ella J. Gehrke, MD; [Arlene V. Drack, MD](#)

Photographer: Douglas B. Critser, CRA, OCT-C, FOPS

Posted: 02/19/2025

Fundus photographs from the left eye (OS) of a 9-year-old female with molecularly confirmed CLN3-associated Batten disease.



Visual acuity at presentation was 20/40. Initial fundus photo shows a parafoveal hyperpigmentation consistent with bullseye maculopathy. The disc is pink, and there is normal vascular caliber.

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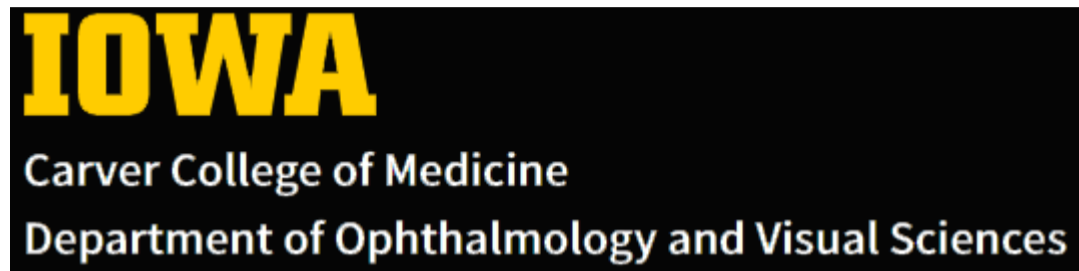
At 5 years after presentation (age 14), visual acuity was 20/800, accounting for the off-center image. The macula has loss of the bullseye appearance, with degenerative hypopigmented appearance to the surrounding retina. There has been interval development of optic nerve pallor and vascular attenuation.

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