

Corneal clouding in Hurler's syndrome

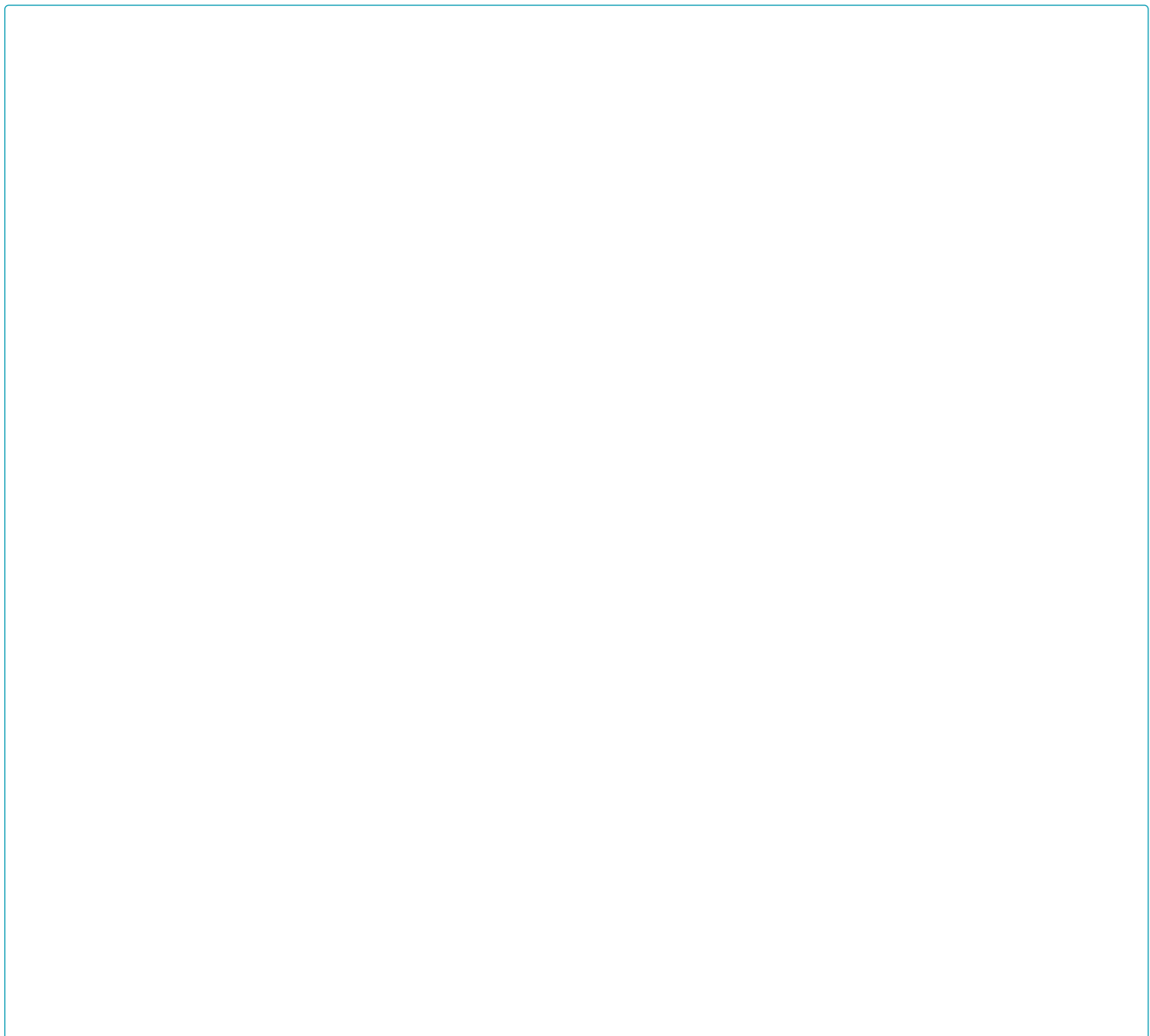
Category(ies): Cornea, Pediatrics

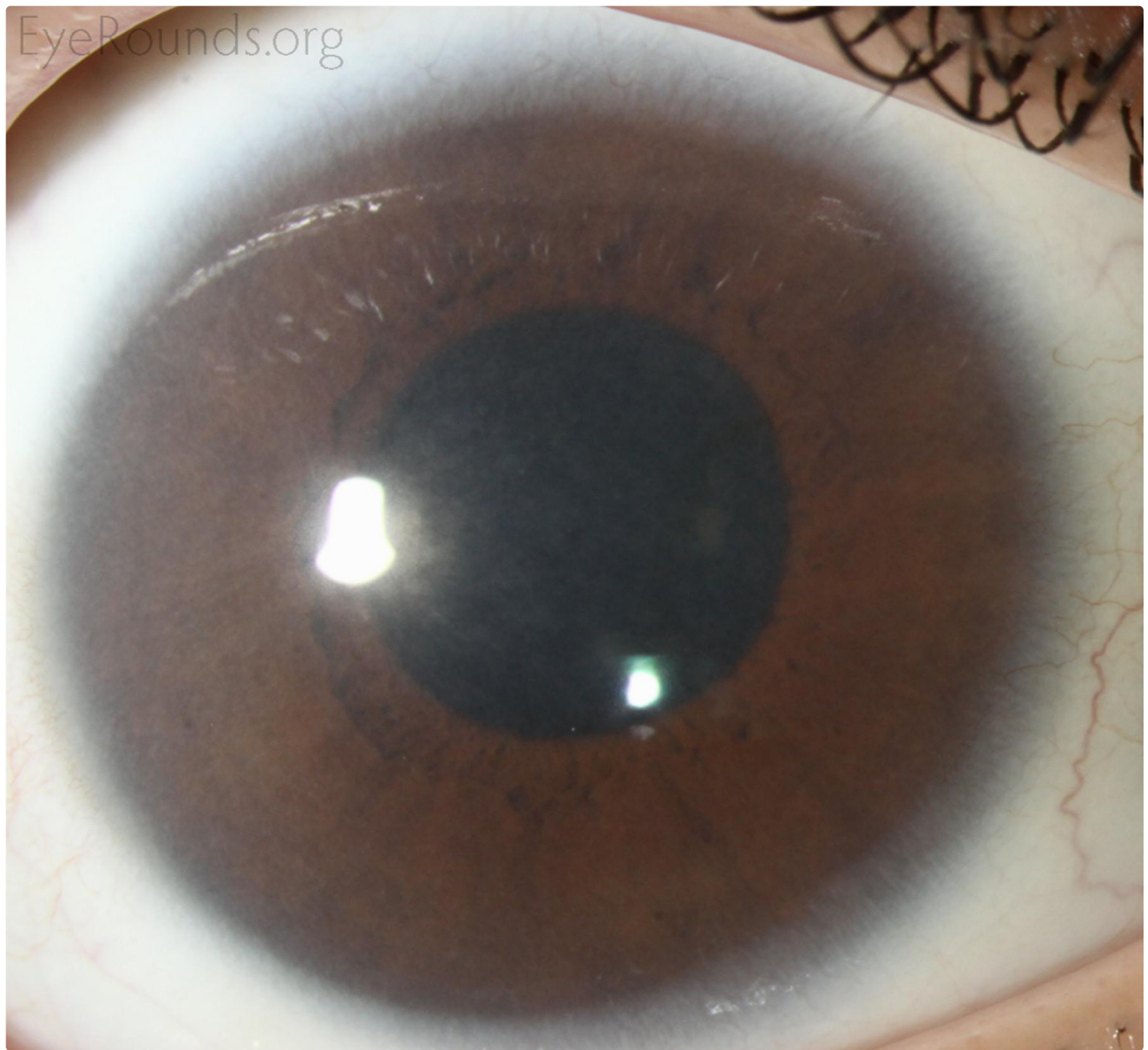
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Photographer: Brice Crister, CRA



Cloudy corneas noted in a 12-year-old patient with Hurler's syndrome, also known as mucopolysaccharidosis type-1 (MPS-I). His alpha-L-iduronidase activity was found to be 0 (normal 0.17 - 0.54). The patient underwent hematopoietic stem cell transplant at the age of 14 months. Corneal clouding is thought to be due to direct infiltration of glycosaminoglycan in the cornea. The corneal opacities first appear in the anterior stroma and are fine, gray, and punctate. Later, the posterior stroma and endothelium become involved. It is felt that the corneal clarity can clear overtime following a hematopoietic stem cell transplant. Retinal degeneration along with optic nerve head swelling and atrophy have also been described in Hurler's Syndrome.

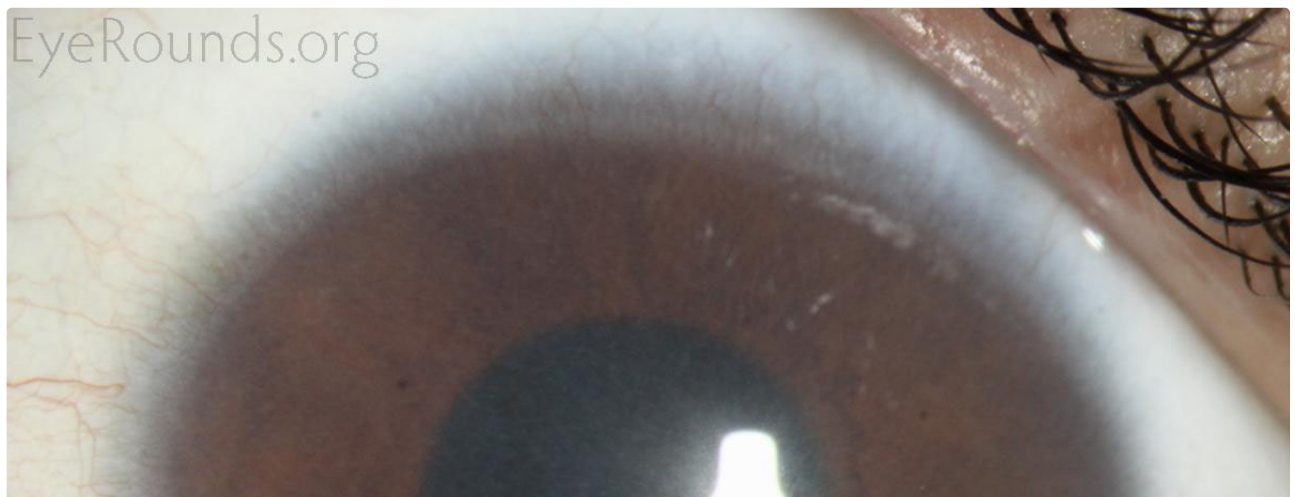


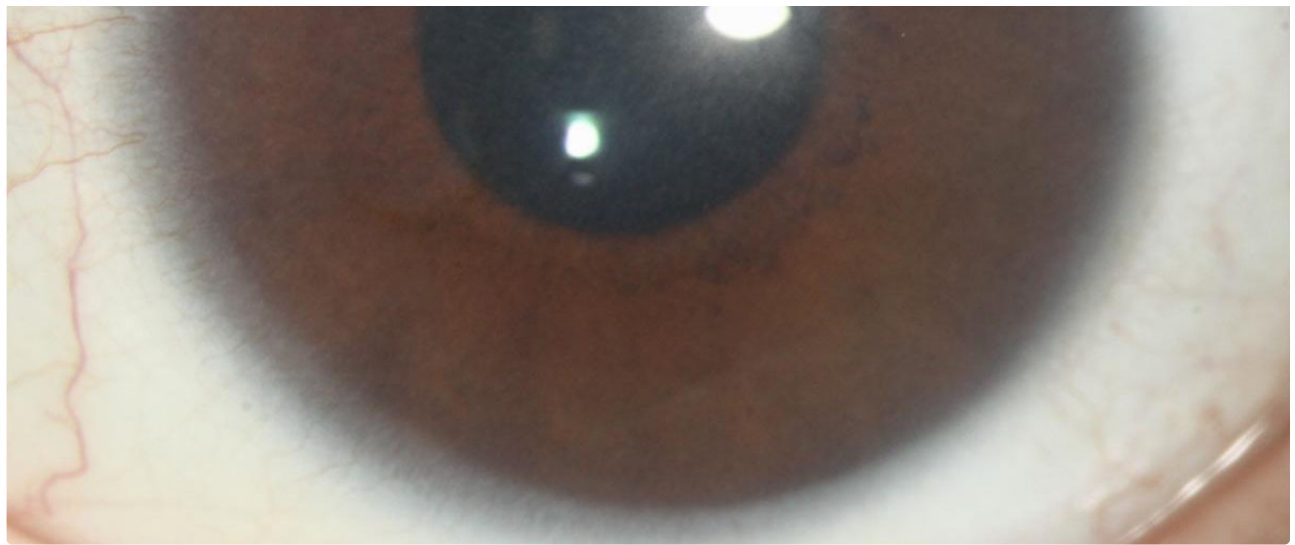


Right Eye

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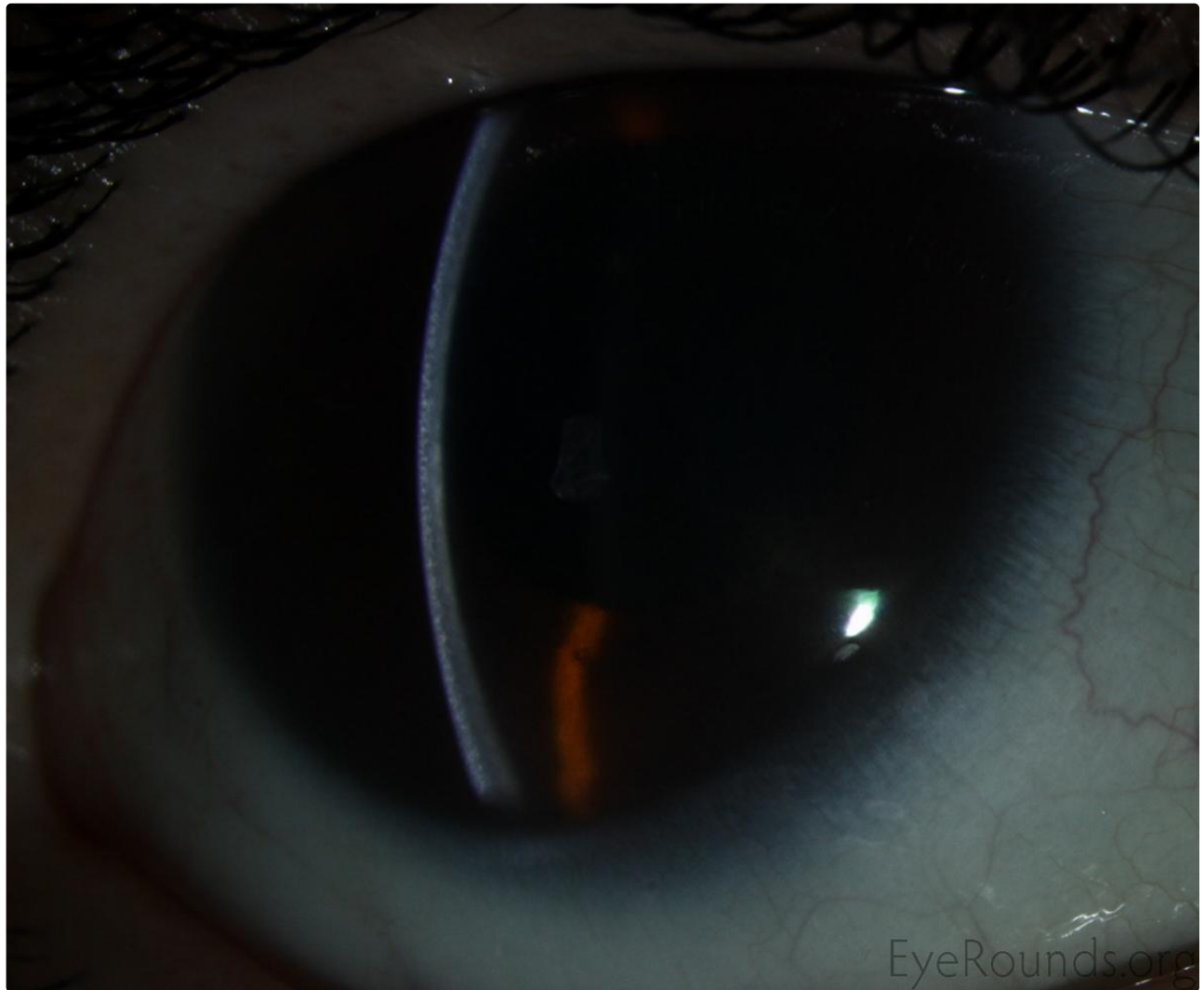




Left Eye

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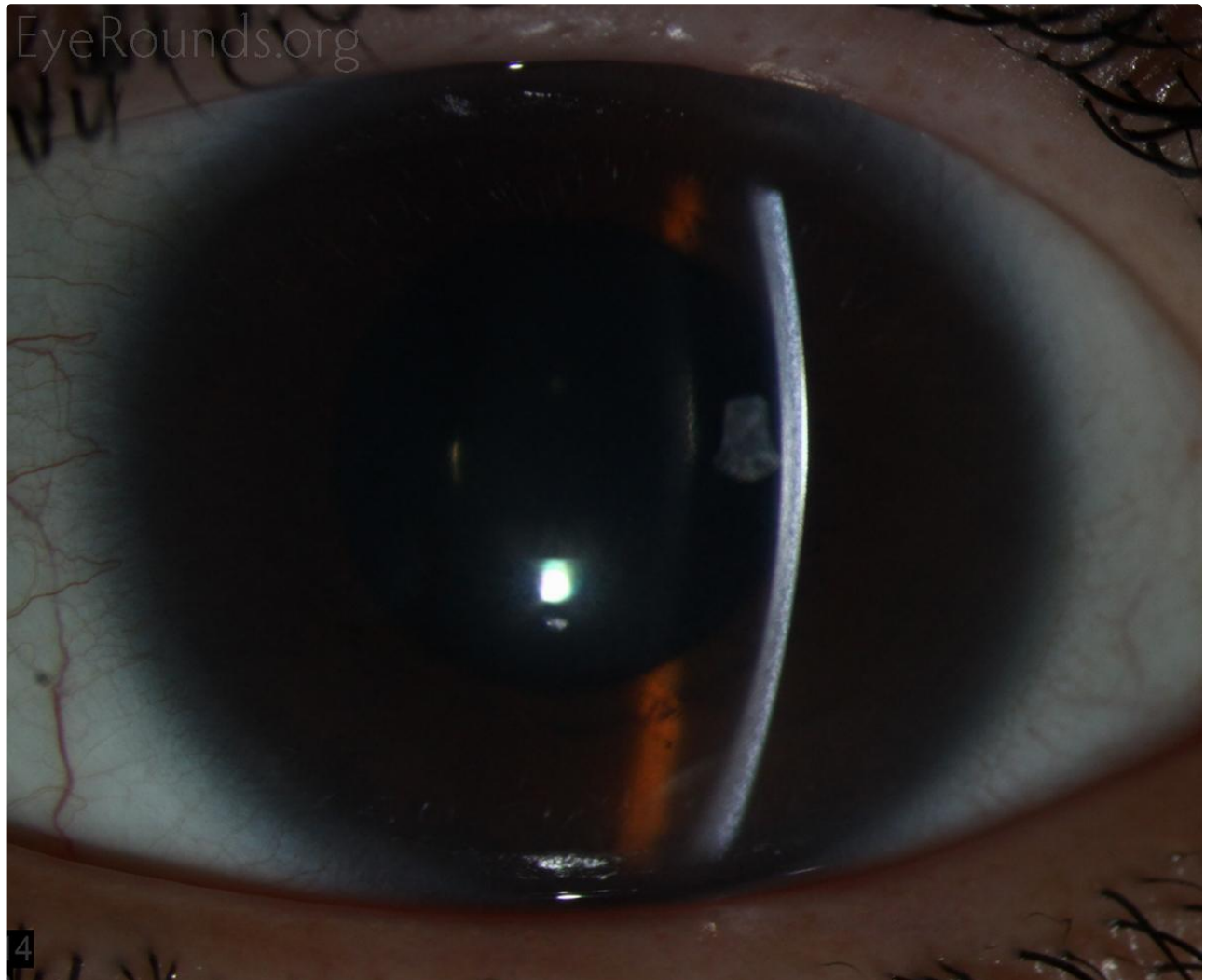
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References:

1. Huang, Y. et al. Ultrastructural study of the cornea in a bone marrow-transplanted Hurler syndrome patient. *Exp. Eye Res.* (1996) 62, 377-387

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