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The role of corneal densitometry in the ophthalmic examination of Fabry's disease, and in the follow-up of the effect of enzyme replacement therapy

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Objective: To study the corneal densitometric values of Fabry's disease with typical corneal vertigo, and to monitor the effect of the enzyme replacement therapy with a Scheimpflug camera

Patients and Methods: The corneal densitometric values of 6 patients with Fabry's disease were compared to the eyes of 5 healthy subjects. In a hemizygotic male patient and heterozygous female patient, 2 studies were performed on both eyes with 2 years follow-up during the therapy. Measurements were performed with Scheimpflug camera Pentacam HR (Oculus Optikgerate) The results were given in standardized grayscale units (GSU).

Results: The mean densitometric value for the entire cornea was significantly higher in the patient group compared to the control group. In the central layer of the corneal surface of 120 μm the mean densitometric values measured in the deepest 60 μm layer were significantly higher in Fabry patients. In the densitometric values of hemizygotic men and heterozygous women, the superficial 120 μm layer was found to be significantly different. In two years of enzyme replacement therapy, the density values for total cornea significantly and a significant decrease in corneal surface 120 μm and the lower 60 μm .

Conclusions: 1. Fabry's disease with cornea verticillatus and subepithelial haze significantly increased densitometric values compared to normal control eyes. 2. Enzyme replacement therapy resulted in a significant decrease in Fabry's disease in corneal densitometry. In the future, the method in examining the efficacy of therapy, as well as in the standardisation of the ophthalmic parameters of the scoring system, can play a significant role.

Biography

Rita Szechey: Besides my other activities (ophthalmologic rehabilitation: vision and eyelid surgeries) I work as a consultant for Fabry -patients at the Budapest Semmelweis University. I am a member of the Fabry Patient Organization and I am committed to patient treatment and the research of the disease.

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