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Efficacy of topical cysteamine in nephropathic cystinosis.

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Abstract

PURPOSE: The aim of this study is to evaluate the efficacy of topical cysteamine 0.55% eye drops in the treatment of corneal cystine crystal deposits in patients with nephropathic cystinosis.

METHODS: Thirty-two patients with nephropathic cystinosis were prospectively included in the study. Patients with corneal cystinosis were treated with topical cysteamine 0.55% eye drops. They were examined before treatment, on each monthly visit and after treatment at the last follow-up. Photophobia was classified as grade 0 (none) for no photophobia, grade 1 (mild) for photophobia in bright light, grade 2 (moderate) for photophobia in room light and grade 3 (severe) for photophobia in dim light. Corneal cystine crystals were graded as grade 0=none, grade 1=1-10 crystals/mm², grade 2=11-50 crystals/mm², grade 3=more than 50 crystals/mm². The main outcome measure was evaluation of photophobia and resolution of corneal cystine crystals.

RESULTS: There were 13 male and 19 female patients. The mean age was 8 years with an age range of 8 months to 19 years. The mean follow-up period was 4.1 years with a range of 2-8 years. Improvement of photophobia was not clinically significant in symptomatic patients. Patients displayed statistically significant worsening of corneal cystine deposits during the follow-up period.

CONCLUSIONS: This study has shown that topical 0.55% cysteamine eye drops may have limited effects in decreasing the corneal cystine deposits in patients with severe forms of nephropathic cystinosis.

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Conflict of interest statement

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