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CATARACT DEVELOPMENT ASSOCIATED WITH LONG-TERM GLUCOCORTICOID THERAPY IN DUCHENNE MUSCULAR DYSTROPHY PATIENTS

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To the Editor: We read with interest the recent article by Rice and colleagues¹ reporting on 596 patients with Duchenne muscular dystrophy (DMD) treated with steroid and evaluated for cataract development. The authors reported that the prevalence of cataracts was 25.1% in patients receiving deflazacort compared with 12.6% of those on prednisone. Only 7 patients (1.4%) required cataract surgery at a mean age of 16.9 years, and all of them were on deflazacort. Although, as pointed out by the authors, this suggests a link between steroid administration and cataract development, a potential influence of the position of the DMD gene mutation cannot be excluded.

Correlations between ocular phenotypes and position of the mutation have been previously suggested. First, in regard to the retina, abnormal dark-adapted electroretinograms (ERGs) were reported to be more frequent in DMD and Becker muscular dystrophy patients with more distal mutations.² More recently, it is the prevalence of red-green color defect among DMD patients that was reported to be associated with deletion downstream of exon 30.³ Relative to the ocular lens, position study in DMD gene mutant and transgenic mice revealed a differential cataract prevalence in mice lacking the main product (mdx)⁴ or the more distal one (Dp71-null mice), with, for the latter, progressive cataract due to loss of lens fibers' organization.⁵ This particularly critical role of Dp71 in the lens is consistent with the relative roles of distal dystrophins isoforms in human retina.

Based on the existing literature, it would be important, first, to determine precisely retinal function (color vision, electroretinography, retinal optical coherence tomography) and

genotype of the 7 patients who required cataract surgery; and, second, to identify patients with deletion downstream of exon 30 of dystrophin gene in patients receiving deflazacort group and those on prednisone.

Such complementary studies could indeed identify a subgroup of DMD patients for whom deflazacort is a risk factor for surgery-requiring cataract formation. Additionally, an association similar to that with ERG could also establish that, in absence of full genotyping, retinal function assessment in DMD patients could be a viable option to identify those patients at risk of complication associated with long-term glucocorticoid treatment.

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