Quantification of Macular Carotenoids Using Autofluorescence Imaging in Patients With Photosensitive Migraine and Benign Essential Blepharospasm

Photophobia describes light sensitivity or abnormal intolerance of light. Patients with photophobia avoid light because of pain or discomfort. Photophobia may be reported by individuals with conditions not necessarily affecting the eye. Two such conditions are migraine and benign essential blepharospasm.1,2 We have previously demonstrated that patients with migraine or blepharospasm are more light sensitive than age-matched control subjects.3

The pathophysiology of light sensitivity is unknown. We suspected that patients with photophobia might have abnormally low levels of the xanthophyll carotenoids lutein and zeaxanthin in their retinas. These carotenoids filter out phototoxic short-wavelength visible light and function as antioxidants.4 We have previously demonstrated that a rose-colored spectacle tint, FL-41, can ameliorate symptoms of light sensitivity in patients with blepharospasm.5 Because of the similarity between the transmission spectra of FL-41 and carotenoids, we hypothesized that photophobic individuals may have reduced levels of carotenoids in their maculae.

Methods. We noninvasively measured macular carotenoid levels in subjects with migraine, subjects with blepharospasm, and 2 independent, age- and sex-matched control groups. Institutional review board approval was obtained, and all subjects provided informed consent. Patients with migraine and blepharospasm were included only if they reported light sensitivity when headache or blepharospasm free. Subjects with both migraine and blepharospasm were excluded. We excluded subjects with any diseases of the macula, with visually significant cataracts, or with best-corrected visual acuities less than 20/80. We excluded subjects who consumed a diet or supplements containing relatively large amounts of carotenoids.

We have developed a novel, light-emitting diode–based, charge-coupled fundus imaging device that uses retinal autofluorescence to quantify and map the distribution of the macular pigments.6 Retinal lipofuscin chromophores naturally autofluoresce. This autofluorescence is reduced at retinal locations containing lutein and zeaxanthin because these carotenoids have a competing absorption spectrum. Once an autofluorescent image is obtained, the relative intensity of the macula is compared with that of the periphery. This analysis yields a pseudo–color image showing the topographic distribution of pigment throughout the macula (Figure). Peak macular pigment optical density (MPOD), or density units, typically encountered in the center of the human macula range from 0 to 1. A 2-tailed t test was used to determine whether differences between subject and control groups were statistically significant.

Results. Demographic characteristics and average MPOD data for the patients with migraine are shown in Table 1. The mean (SD) peak MPOD for the 21 patients with migraine was 0.34 (0.15). The mean (SD) MPOD for the 21 control subjects was 0.21 (0.13) (P = .006).

Demographic characteristics and average MPOD data for the patients with blepharospasm are shown in Table 2. The mean (SD) MPOD for the 16 patients with benign essential blepharospasm was 0.14 (0.12). The mean (SD) MPOD for the corresponding 16 control subjects was 0.20 (0.13) (P = .16).

Figure. A typical autofluorescent image (A) and a false-color topographic map (B) of the distribution of pigment in a human macula. The instrument detects fluorescence from retinal lipofuscin chromophores (autofluorescence) and thereby indirectly quantifies and spatially images the distribution of macular pigment. The lipofuscin fluorescence intensity is reduced at all retinal locations containing pigment because these pigments have a competing absorption spectrum. The distribution of macular pigments is spatially mapped by projecting the autofluorescent wavelength onto the macula and its periphery and comparing lipofuscin fluorescence intensities at each location. A false-color topographic image map is generated, showing the magnitude and spatial distribution of macular pigments. In this subject, red indicates that pigments are most densely concentrated in the central macula. This image was obtained from a control subject in this study.
Comment. Contrary to our original hypothesis, we did not find reduced levels of macular pigments in our light-sensitive subjects. Instead, we found that subjects with migraine had significantly higher levels of macular pigments compared with a control group. Although patients with blepharospasm had lower levels of macular pigments, this difference was not statistically significant.

It is not possible to tell from our experiments why migraineurs with chronic light sensitivity accumulate higher levels of macular pigment or whether this accumulation is clinically significant. It is possible that the macula accumulates these compounds in an effort to mitigate light sensitivity. The mechanisms that underlie carotenoid absorption, protein binding, transport, and storage are highly complex. There are numerous points in these processes where carotenoid metabolism could be affected in some light-sensitive individuals.

In conclusion, a deficiency of macular pigments does not appear to be involved in the pathogenesis of photophobia in patients with light sensitivity. Further research will need to be completed to elucidate the pathophysiology of photophobia.

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Author Contributions: All authors had full access to all of the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Financial Disclosure: None reported.

Funding/Support: This work was supported by grant HL07744 from the National Institute of Diabetes and Digestive and Kidney Diseases (Mr Frandsen and Ms Llop), grant EY11600 from the National Institutes of Health (Dr Bernstein), and an unrestricted grant to the Department of Ophthalmology and Visual Sciences, University of Utah Health Sciences Center, from Research to Prevent Blindness.

Previous Presentation: This paper was presented at the 36th Annual Meeting of the North American Neuro-Ophthalmology Society; March 9, 2010; Tucson, Arizona.

Rapid Formation and Resolution of Cataracts Following Orthopedic Surgery for a Patient With Charcot-Marie-Tooth Disease

A 45-year-old man was referred for a third opinion regarding vision loss. The vision loss began 2 days after a 4-hour, unremarkable foot surgery for a foot deformity due to type 2 CMTD, an autosomal dominant primary axonal neuropathy. The patient’s medical history was otherwise unremarkable. Preoperative 7-item basic metabolic panel and complete blood cell count findings were unremarkable. Postoperatively, his blood chem-

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<td>0.34 (0.15)</td>
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Abbreviation: MPOD, macular pigment optical density.

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Abbreviation: MPOD, macular pigment optical density.


Rapid development and nonsurgical resolution of significant cataracts is extremely rare. Herein, we report an unusual case of bilateral, rapidly developing cataracts following orthopedic surgery for a patient with Charcot-Marie-Tooth disease (CMTD). The cataracts regressed within 45 days of surgery.

Report of a Case. A 45-year-old man was referred for a third opinion regarding vision loss. The vision loss began 2 days after a 4-hour, unremarkable foot surgery for a foot deformity due to type 2 CMTD, an autosomal dominant primary axonal neuropathy. The patient’s medical history was otherwise unremarkable. Preoperative 7-item basic metabolic panel and complete blood cell count findings were unremarkable. Postoperatively, his blood chem-