addition, the duration of treatment to induce retinal reattachment is currently unknown. However, patients with IH have been treated for several months.

Hemangiomas consist histologically of cavernous and capillary vascular networks. The mechanism by which oral propranolol aids in the resolution of exudative retinal detachment in DCH associated with Sturge-Weber syndrome is unknown. It is possible that, similar to IH, there is vasoconstriction of the DCH due to decreased release of nitric oxide, blocking of proangiogenic signals including vascular endothelial growth factor and basic fibroblast growth factor, and apoptosis in proliferating endothelial cells with vascular tumor regression.

To our knowledge, the benefits of propranolol therapy have not been reported in adult hemangioma or for DCH. This is the first reported case of propranolol treatment in an adult with exudative retinal detachment in DCH associated with Sturge-Weber syndrome.

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Financial Disclosure: None reported.

Funding/Support: This work was supported in part by the Arevalo-Coutinho Foundation for Research in Ophthalmology, Caracas, Venezuela.

Previous Presentation: This paper was presented at the 34th Annual Meeting of the Macula Society; March 9, 2011; Boca Raton, Florida.


Oguchi Disease With Unusual Findings Associated With a Heterozygous Mutation in the SAG Gene

Oguchi disease is a type of congenital stationary night blindness with an autosomal recessive inheritance pattern. Two causative genes have been reported for Oguchi disease: the SAG and GRK1 genes. Homozygous Oguchi disease is characterized by a golden-yellow discoloration of the fundus that disappears after prolonged dark adaptation, called the Mizuo-Nakamura phenomenon. The International Society for Clinical Electrophysiology of Vision—protocol bright-flash electroretinograms (ERGs), performed after 30 minutes of dark adaptation, are typically electronegative with a severely reduced b-wave and milder reduction of the a-wave.1 After 3 to 4 hours of dark adaptation, both amplitudes recover to nearly normal, especially the a-wave.2 However, the recovered rod function is rapidly lost after a short light exposure or a single bright white flash.3

We describe a case of Oguchi disease with unusual findings caused by a putative heterozygous mutation in the SAG gene.

Report of a Case. A 40-year-old woman with visual acuity of 20/20 OU had fundus abnormalities and was referred to our institute. She had photophobia but did not report night blindness. There were no autosomal dominant family history. The retina had a golden-yellow appearance (Figure, A). The Mizuo-Nakamura phenomenon was observed after 30 minutes of dark adaptation (Figure, B). Sequencing of the SAG gene identified a heterozygous mutation of 1147delA at codon 309. No mutation was found in GRK1.

The International Society for Clinical Electrophysiology of Vision protocol was used to record the ERGs. The scotopic ERGs after 30 minutes of dark adaptation showed slightly reduced amplitude and delayed implicit time in b-wave (Figure, C). The bright-flash ERG (30 candelas-seconds/m²) had a positive configuration, although the b:a ratio was lower than normal (Figure, C). The photopic and flicker ERGs performed after 10 minutes of light adaptation were normal (Figure, C). To determine the extent of the rod function recovery, bright-flash ERGs were recorded 4 times at 30-second intervals after 30 minutes of dark adaptation. During the 4 stimuli, the waveform changed from the positive pattern to a negative configuration with a severely reduced b-wave and additional milder reduction of the a-wave, which is characteristic of homozygous Oguchi disease (Figure, D). To our knowledge, this phenomenon has never been reported in normal eyes, in eyes with the typical type of Oguchi disease, or in other cases of Oguchi disease with the same heterozygous SAG mutation (Figure, D). The superimposed ERGs elicited by the 4 consecutive flashes show the variation of rod function recovery (Figure, E).

Comment. To our knowledge, this is the first case of Oguchi disease with a distinct fundus appearance and mild electrophysiological abnormalities associated with a putative heterozygous SAG mutation. However, we cannot exclude the possibility that another mutation exists in the intron of another allele, which causes the mild phenotype in this patient.

The repetitive-flash ERG protocol was crucial for the diagnosis. It has been reported that double- or triple-flash stimulations after prolonged dark adaptation induce ERG alterations in typical patients with Oguchi disease.3 However, the use of a 30-second interval allowed us to follow the degree of rod function recovery.
Arrestin and rhodopsin kinase act in sequence to deactivate rhodopsin to stop the phototransduction cascade. Results of molecular biological studies have suggested that residual arrestin activity correlates with the severity of the clinical phenotype. However, in our case it was more difficult to determine the relationship between the putative heterozygous mutation of the SAG gene and the mild electrophysiological abnormalities in the rod function recovery. A modifying effect of deactivating rhodopsin should be considered.

The time required for the reappearance of the rod function demonstrated in the electrophysiological study and the time required to demonstrate the Mizuo-Nakamura phenomenon were nearly identical. We suggest that the physiological basis for the Mizuo-Nakamura phenomenon may be closely related to the abnormal deactivation of rhodopsin.

**Figure.** Fundus photographs showing the Mizuo-Nakamura phenomenon before (A) and after (B) 30 minutes of dark adaptation. C, The electroretinograms (ERGs) recorded according to the International Society for Clinical Electrophysiology of Vision protocol. D, The ERGs elicited by 4 repetitive flashes at interstimulus intervals of 30 seconds. E, Superimposed ERGs elicited by 4 flashes. The ERGs are from our patient with Oguchi disease (case), another patient with Oguchi disease with a heterozygous mutation (carrier), a typical patient with Oguchi disease, and a healthy subject. DA indicates dark adaptation; LA, light adaptation.

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**Financial Disclosure:** None reported.