Spectral thresholds in macular degeneration

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SUMMARY Spectral sensitivities were measured in 18 normal eyes, 9 eyes in patients with senile macular degeneration, 4 patients with Stargardt's juvenile macular degeneration (JMD), and 2 patients without conclusive signs—that is, genetic or morphological abnormalities—to indicate the cause of loss of central vision. Spectral sensitivity, testing for which included measurements on white, yellow, purple, and blue backgrounds, is here used as an aid in differential diagnosis for cases of macular degeneration.

Macular degeneration may be classified by the age of onset of symptoms, fundus appearance, and/or hereditary patterns. The measurement of spectral threshold may be used in the categorisation of the degeneration as an aid in differential diagnosis. Although there is no adequate treatment for it, distinguishing between the juvenile macular degenerations and senile macular degeneration may have importance in predicting the course of loss of visual acuity as well in understanding the hereditary factors for the purpose of genetic counselling.

In macular degeneration the initial presenting symptom is a loss of central visual acuity over the course of a few months to a year. Routine testing on our patients includes ophthalmoscopy, visual fields (including an Amsler grid test), fluorescein angiography, electroretinography (ERG), electro-oculography (EOG), and visually evoked potentials (VEP). Lack of positive results in any of these tests in a young person might indicate malingering.

Indication of a central vision defect on the Amsler chart in a patient in his second or third decade without other signs or family history of macular degeneration may leave one in doubt as to the source of the vision loss. Spectral sensitivity curves measured in patients with central vision losses down to visual acuity at 6/60 may show a uniquely high sensitivity in the blue region as compared with the sensitivity to spectral lights in the green to red range—for example in Stargardt's juvenile macular degeneration (atrophic fundus flavimaculatus).^{1,2} In some young patients malingering may be detectable by the shape and

reproducibility of the spectral threshold settings (we have satisfactorily measured children as young as 7 years old on the apparatus at the Manchester Royal Eye Hospital).

Methods

The methods used are described by Alvarez *et al.*³ The subject's task was to adjust the intensity of a 1°, 0.5 second test flash until it was just visible. In addition to the 1000 trolonds (td) white background used in the previous paper, the following 3 coloured background lights were used in some experiments: blue, Ilford filter 305, intensity 1000 td; purple, Kodak filters no. 34 plus two CC50R, 1000 td; yellow, Schott OG530, 10000 td.

Results

A normal spectral threshold (average for 18 eyes) is shown in Fig. 1a for 0.5 second flashes on a white background. The peak in the blue region of the spectrum near 440 nm has about the same sensitivity as the 2 peaks at longer wavelengths. The height of this blue peak relative to the peaks at longer wavelengths changes in characteristic ways in macular degeneration.

Patient 1. A 52-year-old male reported difficulty in reading; visual acuities were 6/24 right and 6/18 left. Ophthalmoscopy revealed a small macular haemorrhage in the right eye and other signs of senile macular degeneration (SMD) in both eyes, including mottled diffuse pigmentation with the associated dystrophy of the macula. No lenticular abnormality

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Fig. 1 Spectral sensitivity for a 1° , 0.5 second test spot on a 1000 td white background derives from: (a) an average of 18 normal eyes; (b) patient 1, a 52-year-old male diagnosed as having SMD; (c) patient 2, an 18-year-old female with diagnosis of Stargardt's JMD.

was noted in this early SMD case. The Amsler grid showed paracentral distortion in both eyes. Fig. 1b is a spectral sensitivity curve for this patient's right eye. Loss of sensitivity was similar to the spectral sensitivity average of 9 eyes with diagnosis of SMD.

Patient 2. An 18-year-old female reported a gradual loss of vision over the past 9 years; acuity was 6/36 in each eye. The fundus showed discrete lesions and some pigment clumping at the macula. ERG and EOG were normal. Flash VEPs were normal, while pattern VEP showed no response. Fluorescein angiography facilitated identification of macular lesions in both eyes. Feathery hyperfluorescent spots at the maculas, symmetrical in the 2 eyes, were typical of the commonest type of atrophic macular degeneration, Stargardt's disease.¹² The profile of her spectral sensitivity curve (Fig. 1c) was very different from either the average normal or that of patient 1 (SMD). Sensitivity in the red/green area of the spectrum was very depressed (1.2 log units of sensitivity) relative to

normal, while the detection of a blue test on a white background seemed to have been spared. The results of the 100-hue test for the right eye are shown in Fig. 2. There was a large error score indicating a colour defect was present. However, there was no determinable axis to identify the type of colour defect or to distinguish this central vision defect from any other in relation to the unique response measured by spectral sensitivities.

A summary of sensitivities for 5 eyes with other diagnostic evidence of JMD is shown in Fig. 3. An average normal spectral sensitivity is included (Fig. 3a) for comparison. The diagnosis of JMD was confirmed by family history, early age of onset, and/or by fluorescein angiography in these cases. No central scotoma was noted on the Amsler grid, and fixation was checked with a projection ophthalmoscope (fixation was unsteady in some patients but there was no gross eccentric fixation).

Patient 3. An 18-year-old female had reduction in visual acuity (approximately 6/60 right and left) over the last 2 to 3 years. The vision was subjectively worse in bright daylight. There was no family history of any eye disease. The fundus appeared normal. The ERG, EOG, and flash VEPs were also normal. Spectral sensitivities for 0.5 second flashes on a white background (Fig. 4a) gave the notable profile (similar to that of the patients in Fig. 3) of a large drop in the red/green sensitivity while preserving a normal threshold for blue. This response is unlike the loss in spectral sensitivities noted for other diseases^{3,4} and similar to that of the diagnosed JMD patients (Figs. 1 and 3).

Patient 4. A drop in visual acuity from 6/5 to 6/12 over a 2-year period was noted in a 27-year-old female. Her general health was good and there was no family history of any eye disorders. Fundus, ERG, and VEPs were normal. Spectral sensitivity on a white background (Fig. 4b) showed a depressed response to the test flash throughout the spectrum (compared with the normal in Fig. 4a), unlike either the SMD or the JMD curves. By means of Marré's⁵ methods of applying a coloured background to

Fig. 2 Results of the Farnsworth-Munsell 100-hue test for patient 2, OD.

Fig. 3 Spectral sensitivities for: (a) mean of 18 normal eyes; (b-f) patients diagnosed as having central vision affected by atrophic fundus flavimaculatus, as measured for a 1° , 0.5 second spectral test spot on a 1000 td white background. Figs. b and c are for right and left eye of a 22-year-old male, d is for the left eye of a 9-year-old female, and e and f are for right and left eyes of a 16-year-old male.

distinguish cone function the spectral sensitivity was determined (Fig. 5b). A large (about 0.8 log units) depression of sensitivity was seen for the blue and purple backgrounds. This indicated a reduction in sensitivity of the red and green sensitive cone mechanisms. However, the spectral sensitivity peak on the yellow background was of normal height (compared Figs. 5a and 5b), which indicated that the blue sensitive cones were spared, as in the preceding cases of JMD.

Discussion

Colour vision deficiencies associated with juvenile macular degeneration may provide examples of type 1 defects according to Verriest's⁶ classification. In type 1 defects there is generally a large loss of foveal

photopic sensitivity, leading to nearly achromatic and scotopic vision.¹⁷ The spectral sensitivity measurements of Hansen⁸ show, however, that there may be a selective sparing of the responses from blue-sensitive cones. The present results show that this is a common finding in JMD (Figs. 1c, 3, 4, and 5). The continuing function of the blue-sensitive cones in JMD may not be evident except by applying special psychophysical tests such as the ones used in this study. This is because the blue cones, like the rods, respond poorly to contrast, flicker, and fine details,⁹ so they do not add much to the overall psychophysical performance expected in measurements of scotopic vision.

In the absence of other signs to indicate the cause of loss of central vision spectral sensitivity measurement may lend weight to the diagnosis of JMD. Patient 3

Fig. 4 Spectral sensitivities for 1° , 0.5 second flashes on a 1000 td white background in cases of difficult diagnosis for vision loss in: (a) patient 3, an 18-year-old female, VA 6/60; (b) patient 4, a 27-year-old female, VA 6/12.

Fig. 5 Spectral sensitivity curves for detecting foveal test flashes on yellow (Y), purple (P), and blue (B) background for: (a) 12 normal subjects; (b) patient 4.

illustrates the type of acquired colour defect (as measured by spectral sensitivity) which may be associated with JMD, though there is an absence of any electrodiagnostic abnormality, the fundus appears normal, and there is no family history of macular degeneration. Patient 4, another case where diagnosis was difficult, especially with the late age of onset, illustrated an overall loss of sensitivity for spectral thresholds (for 0.5 second flashes) on a white background. But the comparative loss of red/green peak sensitivity relative to blue could be clearly demonstrated by a variation of the testing procedure, that is, changing the backgrounds for the test flash to yellow, purple, or blue. The relatively high sensitivity on a yellow background (Fig. 5b) thus indicates a sparing of the output from blue-sensitive cones, which again supports the diagnosis of JMD.

The results of spectral sensitivity measurements are quite different from those of central vision loss due to SMD—for example, in patient 1 described here. The severe loss of blue sensitivity as well as some loss in the longer wavelengths of the spectrum has been noted in all other SMD patients whose spectral sensitivity we have measured.

Loss of central acuity may lead some patients to have unsteady or slightly eccentric fixation. Although using an eccentric fixation point may increase the blue peak sensitivity relative to the green/red range of the spectrum, observed changes with eccentric viewing in the normal were not sufficient to explain the difference seen in the patients here.

CONCLUSION

Macular degenerations may be classified by age of onset, hereditary pattern, and/or by some of the routine tests previously mentioned. The ability to diagnose a sudden loss of visual acuity as a primary atrophic macular degeneration of a type first

described by Stargardt may be important in (1) genetic counselling and in a search for afflicted family members; (2) prediction of the course of the disease (the visual acuity rarely progresses to worse than 6/60 in this type of JMD¹); (3) distinguishing a JMD patient from another cause of loss of central acuity, including malingering.

The measurement of a spectral sensitivity may aid in the diagnosis of JMD. The spectral thresholds measured can vary with ocular disease as discussed in the previous article, but only in JMD is blue sensitivity spared while considerable depression may be observed in green/red sensitivity.

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