Incremental Light Detection Thresholds Across the Central Visual Field of Human Albinos

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**Purpose.** The authors investigated central retinal function in albinism by measuring incremental light detection thresholds in a group of oculocutaneous human albinos.

**Methods.** Eleven oculocutaneous human albinos (six tyrosinase negative and five oculocutaneous positive), six patients with idiopathic congenital nystagmus, and six normal control subjects participated in the study. Using a Goldmann bowl perimeter, incremental light detection thresholds were measured in the vertical meridian across the central ± 30° of the retina. Target presentation times were 1 sec for all subjects, and in the case of four albinos and one patient with idiopathic nystagmus, they were limited to the low-velocity period of each nystagmus cycle.

**Results.** For the normal control subjects, the maximum sensitivity was found to be −0.60 ± 0.10 log units. By comparison, at 0°, a range of sensitivities was obtained from the albinos subjects (−0.9 to −2.1 log units) and from those with idiopathic nystagmus (−0.7 to −1.9 log units). The albinos had diverse retinal sensitivity profiles ranging from a near-normal peaked curve to a flat homogeneous profile. A variety of sensitivity profiles was also detected in those with idiopathic nystagmus, although, compared with the albinos curves, a greater proportion were peaked. No sensitivity differences were found between the short and the longer target presentations.

**Conclusions.** The variety of retinal sensitivity profiles obtained in this study suggests that, in albinism, considerable intersubject variability in the degree of foveal hypoplasia exists and that albinos “foveal” function can reach near-normal levels, for at least some visual tasks. Invest Ophthalmol Vis Sci. 1993;34:1683–1690.

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One of the principal ocular features of all forms of albinism is foveal hypoplasia. First described at the turn of this century, the absence of normal foveal differentiation has been confirmed by histologic evidence from both human and primate eyes. These studies have reported that, in albinos, the ganglion cell layer is present throughout the retina, there is no rod-free area, and there is an absence of a foveal pit. In addition, the presence of congenital nystagmus, high refractive errors and intraocular light scatter all adversely affect retinal image quality. Thus, visual acuity in albinism typically ranges from 3/60–6/12. Currently, comparatively few studies have investigated the functional capacity of the albinos central retina. In one, the visual fields of a group of albinos were examined using kinetic perimetry, and it was found that all 16 subjects had a relative or absolute central scotoma. This was attributed to foveal hypoplasia. Thirty years later, others investigated the visual fields of 13 albinos and found that 9 had contracted...
fields. More recently, psychophysical studies have shown that the major factor limiting albinos spatial performance is foveal hypoplasia. Furthermore, on the basis of their studies, the latter group proposed that the central retina in an adult albinos had many anatomic and functional similarities with that of a normal 10-month-old infant.

Evidence from experiments using the simultaneous monitoring of eye and retinal image movements have indicated that some albinos have a consistent fixation pattern in which the low-velocity period of the nystagmus cycle regularly coincides with a central retinal site. Such precise foveation strategies suggest that there may be some advantage in using the central retina over other retinal locations. The aim of this study was to investigate central retinal function in albinos by measuring incremental light thresholds within ±30° of the center in a group of oculocutaneous albinos.

MATERIALS AND METHODS

Incremental light thresholds were determined with a Goldmann 940 ST hemispheric projection perimeter (radius, 300 mm). The background luminance was set at 10 cd/m², and the test target luminance was varied in 0.1-log unit steps using neutral-density filters. The maximum and minimum target luminances were 318 and 0.318 cd/m², respectively. Each session commenced with a 5-min adaptation period, and if large eye movements were observed, fixation was monitored with the built-in telescope, and if large eye movements were observed, threshold measurements were repeated.

The subject population consisted of 6 normal control subjects (age range, 18–24 yr), 6 patients with congenital idiopathic nystagmus, and 11 oculocutaneous albinos. These subjects were taken from a larger subject pool (n > 100) and had all previously undergone a full optometric assessment (including ophthalmoscopy and color vision testing) to exclude the presence of other ocular disorders associated with congenital nystagmus, such as aniridia and achromatopsia. Biochemical tests and clinical examination, in combination with the details of family history and personal tan-

<table>
<thead>
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<th>TABLE I. Clinical Data for the 11 Albino and 6 Idiopath Subjects</th>
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Idio = congenital idiopathic nystagmus, TNOCA = tyrosinase-negative oculocutaneous albinism, TPOCA = tyrosinase-positive oculocutaneous albinism, PAN = periodic alternating nystagmus. Nystagmus waveforms were: Bidirectional (BD), Dual jerk (DJ), Pure jerk (J), Jerk with decreasing velocity (Jdv), Jerk with extended foveation (Jef), Pure pendular (P) and Pseudocycloid (PC). * = data not available.

Retinal sensitivity thresholds were determined for Target 1 (6.5’ of arc in diameter) presented along the vertical meridian within ±30° of the center. From 0–20°, measurements were taken at 2° intervals and between 20–30°, at 5° intervals. For all but the central measurements, the subjects were instructed to fixate a small circular mirror at the center of the perimeter. Foveal testing was done by asking the patient to fixate the center of a diamond pattern of four light spots projected 5° along the horizontal meridian, with the test target being presented at its center. An ascending method of limits was employed to find the threshold. At each target luminance level, the stimulus was presented for approximately 1 sec. Throughout each session, fixation was monitored with the built-in telescope, and if large eye movements were observed, threshold measurements were repeated.

The subject population consisted of 6 normal control subjects (age range, 18–24 yr), 6 patients with congenital idiopathic nystagmus, and 11 oculocutaneous albinos. These subjects were taken from a larger subject pool (n > 100) and had all previously undergone a full optometric assessment (including ophthalmoscopy and color vision testing) to exclude the presence of other ocular disorders associated with congenital nystagmus, such as aniridia and achromatopsia. Biochemical tests and clinical examination, in combination with the details of family history and personal tan-
ning experience, showed that six of the albinos were tyrosinase negative and five were tyrosinase positive.\textsuperscript{19} Bilateral conjugate horizontal nystagmus was exhibited by 4 of those with idiopathic nystagmus and 10 of the albinos. The remaining two subjects had bilateral torsional nystagmus. A summary of the relevant clinical data, including details of each subject’s typical nystagmus in primary gaze, is presented in Table 1. For the five subjects with periodic alternating nystagmus (four albinos and one with idiopathic nystagmus), amplitude and frequency data were calculated using the means of both the right- and left-beating phases.

Five subjects (one with idiopathic nystagmus and four albinos) were re-examined using a shutter device that limited target presentation to the foveation period. This arrangement relied on the registration of horizontal eye movements by an infrared eye position sensor, the differentiated output of which was fed into a Farnell pulse generator. After a preset delay of up to 30 msec, a triggering pulse was sent to a solenoid when the velocity reached a predetermined threshold. Attached to the solenoid was a shutter that interrupted the presentation of the target. When the pulse was generated, the shutter opened, and the target was exposed for a predetermined period of time (40 or 80 msec). The velocity threshold was varied so that a pulse was only generated during each nystagmus fast phase. In this way, the target was exposed during the periods of low retinal image velocity.

Three measures of visual performance were also made. First, monocular visual acuities were recorded using a high-contrast (90%) Bailey-Lovie chart located in the primary position. Visual acuity was recorded as the log of the minimum angle of resolution (log MAR) from 1.0 (Snellen equivalent, 6/60) to 0.0 (Snellen equivalent, 6/6). Second, retinal fixation behavior was assessed by simultaneously recording eye and retinal image movements (by fundus video analysis\textsuperscript{13,20}) or by direct observation with a Canon (Tokyo, Japan) nonmydriatic fundus camera. Third, the distribution of velocities that comprised ten consecutive nystagmus slow phases of each of the idiopathic and albino subjects was computed.\textsuperscript{21}

The tenets of Declaration of Helsinki were followed. Informed consent was obtained from all subjects before each procedure.

**RESULTS**

**Normal Sensitivity Function**

The relationship between log sensitivity and retinal eccentricity for six normal subjects is illustrated in Figure 1. These data were consistent with the results from previous studies showing sensitivity peaks at the fovea (0°) and then decline with distance from the fovea.\textsuperscript{22-24}

![Figure 1. The relationship between log sensitivity and retinal eccentricity for the six control subjects. The dotted lines indicate ± one standard deviation. SUP, superior visual field; INF, inferior visual field.](Downloaded From: http://iovs.arvojournals.org/pdfaccess.ashx?url=/data/journals/iovs/933398/ on 06/06/2017)

The maximum sensitivity was found to be −0.60 ± 0.10 log units, and the mean difference in sensitivity from 0–4° and from 0–10° in the superior and inferior visual fields was +0.50 and +0.75 log units, respectively.

**Albino Sensitivity Function**

The 11 albino subjects showed a great diversity of incremental sensitivities at the 0° location (Figs. 2, 3). By comparison with the normal group, the albino peak sensitivity, which ranged from −0.9 log units for Subject AB (Fig. 2A) to −2.1 log units for Subject AG (Fig. 3E) was reduced. Most of the albinos exhibited sensitivity losses of at least 1.0 log unit (Table 2). Five of the 11 albinos (Figs. 2A–C, E, 3A) exhibited peak sensitivities at 0°. Five showed a small 0.1 log unit depression in sensitivity (Figs. 2D, 3B–E), and Subject TG (Fig. 3F) had a 0.2-log unit drop in sensitivity at 0° compared with the sensitivity at an eccentricity of 2° and/or 4°. Unlike the normal control subjects, the albinos showed retinal sensitivity profiles that qualitatively varied from the peaked profiles to those that were virtually flat. To aid interpretation of these curves, the mean drop in sensitivity from 0–4° and from 0–10° in the superior and inferior visual fields was determined for each subject. The sensitivity ratios (ie, the mean difference in log sensitivity between two retinal locations) ranged from +0.49 to −0.25 log units for the 0–10° field locations. On the basis of these sensitivity ratios, the retinal profiles were categorized into one of three classes. This classification is given in Table 3. Thus, Subjects AB (Fig. 2A) and LA (Fig. 2B) with peaked profiles and high sensitivity ratios belonged to Class I and those with flat profiles (DK, Fig. 3A; GT, Fig. 3B; LR, Fig. 3C; TD, Fig. 3D; AG, Fig. 3E; and
FIGURE 2. The relationship between log sensitivity and retinal eccentricity for five oculocutaneous albinos showing Class I (A) and (B) and Class II (C–E) profiles. SUP, superior visual field; INF, inferior visual field.

TG, Fig. 3F) fell into Class III. The remainder (JG, Fig. 2C; CG, Fig. 2D; and SL, Fig. 2E) were grouped into Class II. Individual sensitivity ratios and other details are presented in Table 2.

Generally, flatter profiles were associated with a lower visual acuity, decreased central retinal sensitivity, and shorter foveation periods. However, there were some marked exceptions. Subject AB (Fig. 2A), who had a small amplitude torsional congenital nystagmus, good monocular visual acuity (0.42 log MAR), and a high central retinal sensitivity (−0.9 log units) displayed a near normal Class I profile. Surprisingly, so did Subject LA (Fig. 2B) who had poor visual acuity (0.94 log MAR), a low central retinal sensitivity (−2.0 log units), and a periodic alternating nystagmus in which an average of only 3.5% of her nystagmus slow phase was spent at velocities of 10°/sec or less. Subject LR (Fig. 3C), who had a visual acuity of 0.92 log MAR, a −1.7 log unit central sensitivity, and a waveform in which 9.3% of the slow phase had a velocity of 10°/sec or less, displayed a flat Class III profile. Unexpectedly, so did Subject DK (Fig. 3A), who had an acuity of 0.62 log MAR, a −1.7 log unit central retinal sensitivity, and a waveform in which 78.2% of the slow phase had a velocity of 10°/sec or less.

The assessment of retinal fixation behavior in the 11 albinos produced diverse results because both accurate and highly variable foveation patterns were observed in each category. The only apparent trend was that a smaller proportion of the albinos that belonged to Class III (one of six) adopted a precise foveation strategy compared with those found in Classes I and II.

Although this study concentrated on retinal sensitivity in the vertical plane, it could be argued that, because all subjects had congenital nystagmus, the presence of continuous retinal image motion may have adversely affected the threshold measurements. This was thought unlikely for two reasons. First, observation of the test eye with the built-in telescope showed that, in most cases, the involuntary oscillations damped down to less than 1° in amplitude during the sensitivity measurement. Second, when target presentation was limited to the foveation periods, by using the shutter device, almost identical profiles were obtained (Figs. 4A–D).

FIGURE 3. The relationship between log sensitivity and retinal eccentricity for six oculocutaneous albinos all showing Class III profiles. SUP, superior visual field; INF, inferior visual field.
Idiopathic Nystagmus Sensitivity Function

Unlike albinos, subjects with idiopathic congenital nystagmus are not considered to have foveal hypoplasia and, therefore, would be expected to show peaked retinal sensitivity profiles. In general, our results confirmed this with four of the six patients with idiopathic nystagmus having Class I profiles (Figs. 5A–D). The major exception was Subject BP (Fig. 5F) who had a low visual acuity of 0.80 log MAR and a markedly reduced central sensitivity of −1.9 log units. Many previous investigations have been done on this subject, and there was no indication that he was an albino. No sensitivity difference was found between target presentations of 1-sec intervals and target presentations limited to the foveation periods (Figs. 4E, 5E).

The distribution of albinos and patients with idiopathic nystagmus across each of the three classes is summarized in Figure 6. Albinos with tyrosinase-negative oculocutaneous albinism appeared in all three classes; however, a relatively large number of tyrosinase-negative albinos (four of six) had flat profiles and were placed in Class III. Although this suggests that “foveal” function and, perhaps, the degree of foveal hypoplasia are linked to retinal melanin levels, firm conclusions cannot be drawn from such a small group.

**TABLE 3. Classification of Retinal Profiles on the Basis of Sensitivity Ratios**

<table>
<thead>
<tr>
<th>Group</th>
<th>Number</th>
<th>$0^\circ$ to $4^\circ$</th>
<th>$0^\circ$ to $10^\circ$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal subjects</td>
<td>6</td>
<td>0.50 ± 0.05</td>
<td>0.75 ± 0.05</td>
</tr>
<tr>
<td>Class I</td>
<td>6</td>
<td>0.49 − 0.25</td>
<td>0.75 − 0.51</td>
</tr>
<tr>
<td>Class II</td>
<td>4</td>
<td>0.25 − 0.10</td>
<td>0.51 − 0.35</td>
</tr>
<tr>
<td>Class III</td>
<td>7</td>
<td>0.10 − 0.25</td>
<td>0.35 − 0.10</td>
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</table>

The normal ratios are the means of six subjects.
matism (> 4.0 D) from an early age, resulting in a greater neural deprivation than is normally encountered in albinism. The fact that Subject DK had consistent but eccentric fixation supports this because no benefit would be gained from foveal fixation if the central retina was homogeneous in sensitivity. Similarly, on the basis of visual acuity and waveform shape, we would have predicted that Subject LA (Fig. 2B) would have exhibited a Class III profile, but a Class I profile was obtained. In this case, the cyclic nature of her periodic alternating nystagmus and, in particular, the great variability in the length of the quiet phases may have led to the discrepancy between the waveform features and profile shape. During perimetry, the quiet phases tended to be of much longer duration than those displayed during eye movement recording sessions.

One of the most surprising results of this study was that, despite the presence of foveal hypoplasia (revealed by ophthalmoscopy), one albino had a near-normal retinal sensitivity profile. However, in this study, only a basic aspect of visual processing, incremental light sensitivity, was assessed. With a more complex psychophysical task, such as form discrimination, the capacity of albino spatial mechanisms may be considerably impaired by comparison with normal abilities. In a recent series of experiments, the spatiotemporal vision of two experienced albino subjects was assessed; one was tyrosinase negative and the other, tyrosinase positive. Both albinos had normal flicker sensitivities; however, their grating and vernier acuities in the center (0°) were approximately fivefold poorer than normal. At 10° in the inferior visual field, the grating and vernier acuities reached normal levels. Our experiments show that only 2 of the 11 albinos had sensitivity ratios (0–10°) comparable with those of the normal control subjects. The others showed reduced sensitivities in the center, the periphery, or both. Thus, unlike the earlier findings, a consistent relationship between albino central and peripheral retinal performance could not be demonstrated. A number of reasons may be proposed for such a difference in the results between our two studies. First, the other group only examined two retinal locations, 0° and 10°. Second, the ongoing congenital nystagmus was largely ignored in their studies. It is well es-

FIGURE 4. The relationship between log sensitivity and retinal eccentricity for four oculocutaneous albinos (A–D) and one patient with idiopathic nystagmus (E) when the target presentation was limited to the foveation periods (the continuous line). The dotted function represents the sensitivity profile for a stimulus presentation of 1 sec. SUP, superior visual field; INF, inferior visual field.

FIGURE 5. The relationship between log sensitivity and retinal eccentricity for six subjects with idiopathic congenital nystagmus. SUP, superior visual field; INF, inferior visual field.
established that variations in the nystagmus waveform shape, intensity, and fixation behavior influence contour detection thresholds.7–9,20,21,27,28 In addition, the two subjects who were evaluated had poor visual acuity (1.0 log MAR), even for albinos. It may be that, with a larger and more representative sample, a variety of retinal detection and resolution profiles would have been obtained.

Finally, our study showed that patients with idiopathic nystagmus also exhibit a variety of retinal sensitivity profiles, and therefore, caution should be exercised when using such patients as control subjects during psychophysical investigations.29

Key Words

albino, incremental light threshold, retinal sensitivity, foveal hypoplasia

References

15. St. John R, Timney B. Sensitivity deficits consistent


