The Photopic Electroretinogram in Congenital Stationary Night Blindness with Myopia

Pierre Lachapelle, John M. Little, and Robert C. Polomeno

Previous studies have reported that subjects affected with congenital stationary night blindness and myopia demonstrated some photopic (cone) abnormalities in their electroretinogram (ERG). By comparing the photopic ERG elicited with a threshold and a suprathreshold stimulus it was found that, at threshold, no significant differences were noted both in the peak time and in the amplitude of ERGs evoked from CSNB and normal subjects. However, a more powerful stimulus (16 times the threshold) yields a significant difference in the ERGs recorded from the two groups. ERGs recorded from CSNB patients are decreased in amplitude with a b-wave peak time that remains normal. First derivative analysis of the ERG wave along with a selective recording of the oscillatory components of the ERG suggest that the only visible anomaly in the suprathreshold photopic ERG of CSNB patients is an absence of the two oscillations normally seen on the ascending portion of the b-wave. Data obtained on normal subjects are also reported that try to explain the functional significance of these two oscillatory potentials. Invest Ophthalmol Vis Sci 24:442-450, 1983

Congenital stationary night blindness (CSNB) with myopia is a condition in which the scotopic system is reported to be abnormal. The association of night blindness and myopia is hereditarily transmitted mainly as an x-linked recessive or autosomal recessive trait. The rhodopsin concentration as well as its kinetics appears to be normal, suggesting that the outer segments of the rods are functioning normally in CSNB. Furthermore, the light- and dark-adapted a-waves are also reported as being normal, suggesting that the photoreceptors (rods and cones) are also normal. The lack of a rod-mediated b-wave has led some investigators to postulate a possible decrease in the cone population or at least an impairment in the photopic (cone) system in CSNB with myopia. CSNB with myopia subjects, however, do not report visual anomalies that could be explained on the basis of a decrease in the cone population. Color vision is reported as being normal or slightly subnormal, and visual acuity is seldomly reported as a major complaint. Finally, the visual fields are normal if performed in the photopic range. However, visual fields measured in the mesopic range are constricted when compared to those obtained in normal subjects. How then can we explain the decrease in the photopic ERG recorded from subjects affected with CSNB and myopia?

In order to reinvestigate this interesting retinal disorder we have studied more closely the morphology of the photopic electroretinogram (ERG) with special attention given to the low voltage, fast oscillation components of the ERG identified as oscillatory potentials. In CSNB with myopia the two oscillations normally recorded on the ascending portion of the b-wave are absent. This finding was also reported by Heckenlively. We believe that at least one of the two missing oscillations (OP3) would be rod dependent. The anomalies in the rod (scotopic) system would explain the absence of that oscillation (OP3) from the photopic ERG and thus contribute to the overall decrease in the photopic ERG. Parts of the present study have been reported elsewhere.

From the Department of Ophthalmology, The Montreal Children's Hospital, McGill University, Montreal, Canada. Supported in part by a grant-in-aid from the McGill University/Montreal Children's Hospital Research Institute. This is publication number 83012 from McGill University/Montreal Children's Hospital Research Institute. Submitted for publication February 16, 1982. Reprint requests: Pierre Lachapelle, PhD, The Montreal Children's Hospital, Department of Ophthalmology, Room D-280, 2300 Tupper Street, Montreal, Quebec, Canada H3H 1P3.

0146-0404/83/0400/442/$1.25 © Association for Research in Vision and Ophthalmology
Materials and Methods

Subject Selection and Preparation

The results reported here were obtained after studying two families in which symptoms of night blindness along with myopia (−5 to −10 diopters [D]) were observed. Fundus examination was found to be within normal limits with the exception of a slight myopic change seen at the disc margin. A similar anomaly was also noted by previous investigators. Visual field testing was performed in one of the patients. Apart from minor field cuts that could account for the astigmatic error of the subject, no other anomalies were found. Color vision evaluated using the Ishihara plates was found to be normal. No clinical anomalies were found that could have been linked with other night blinding conditions such as retinitis pigmentosa, choroideremia, or vitamin A deficiency for example.

Although our prime concern is not to discuss the mode of transmission of the condition, it may be important to describe briefly the two families in that respect. In the first family, the affected members were the grandfather (TD) and the grandson (SJ). The mother (daughter of TD) of SJ was also examined. The only anomaly found was a slight delay in the peak time of her scotopic b-wave. In the second family, both the brother and sister were affected. Their mother and father were examined. Only the father was shown to have slight photopic ERG changes that could represent a mild variant of those found in CSNB subjects. However, the father was not night-blind nor was he myopic. No other members of the two families' pedigree was available for examination making difficult a final diagnosis as to the mode of inheritance. However, the photopic anomalies observed were identical in the two families and, in that respect, a final diagnosis on the mode of transmission will not be of any help in interpreting this photopic anomaly.

The control group consisted of 13 normal subjects whose ERG examinations coincided in time with that of CSNB patients. The subject's eyes were dilated (cyclopentolate hydrochloride 1%, phentolamine hydrochloride 10%) and the cornea topically anesthetized (proparacaine HCl 0.5%). Corneal contact lens (Medical Workshop Inc.) were used to pick up the retinal signal. A reference electrode was placed on the forehead and a ground electrode on the earlobe.

Light Stimuli

The subjects were then placed in front of a full-field stimulator (45 cm in diameter) and light-adapted for 3 min at a background luminance of 50 lumen/m². The cone and rod electroretinograms were separated using a technique previously reported by Brunette. Briefly, after preadapting the subject to a luminous background the photopic portion of the ERG examination was performed with light stimuli delivered by a Grass PS-22 photostimulator. The intensity of the flash stimulus was calculated as ranging from 0.69 cd/m²/sec (setting 1) to 8.70 cd/m²/sec (setting 16). Examples of the photopic ERG were photographed for each of the intensities offered, after which the background adapting light was closed and the subject was dark adapted for 16 min. During that period a neutral density filter was placed in front of the flash to attenuate its intensity by 3.0 log-unit. The rod-mediated ERG was recorded using the same incremental factor offered on the photostimulator.

Recording

Each retinal signal was fed in two distinct sets of preamplifiers (Grass P-511). The first set had an amplification factor of 1000x and a bandwidth fixed between 1–300 cps (ERG). The second set had an amplification factor of 20,000x and a bandwidth fixed between 100–1000 cps (OP). Both signals were then fed in a FM tape recorder (Hewlett Packard 3968A) and kept for further analysis. The electroretinograms were also monitored on an oscilloscope, and a single sweep of the different intensities used were photographed for analysis. The oscillatory potentials recordings were fed in a signal averager (Tracor Northern NS575A). Only two presentations of a given stimulus were used to build the average. The signal thus obtained was reproduced on a X-Y plotter (Hewlett Packard 7015B).

Data Analysis

The peak time of the b-wave was measured from the onset of the stimulus to the peak in the amplitude of the b-wave. The amplitude of the b-wave was measured from the trough of the a-wave to the peak of the b-wave. A delay of about 20 msec was incorporated between the triggering of the oscilloscope (or averager) sweep and the delivering of the stimulus, thus permitting a baseline recording. The first derivative of the photopic ERG wave was also analyzed. The derivative was obtained by computing the changes in voltage per millisecond measured during the first 80 msec after flash onset. The data thus collected reflect the slope changes (μV/msec) occurring during the building of the ERG wave. The ERG real time was then divided into 4-msec bin, and the slope values calculated within that 4-msec time slot were added. Each 4-msec bin was then weighed by comparing the magnitude of the slope change for a given 4-msec bin,
with the total slope changes measured for the 20 4-
millisecond bins that composed the entire ERG wave
analyzed. Processing the data in this particular way
permitted us to quantify the importance of a given
electrical segment to the building of the entire elec-
troretinogram.

Results

At threshold (Fig. 1A) the amplitude of the pho-
topic ERG in normals (80.00 ± 17.43 μV) does not
differ significantly (t = 0.75; df = 15) from that mea-
sured in CSNB patients (77.44 ± 19.44 μV). How-
ever, a more powerful stimulus (16 times the thresh-
hold: Fig. 1B) yields a significant difference (t = 6.13;
df = 15) in the ERG amplitude of normals (228.33
± 39.90 μV) as compared to CSNB patients (135.00
± 32.60 μV). Despite the intensity of the stimulus
used, the peak time of the b-wave in CSNB recordings
remains normal. The amplitude of the photopic su-
prathreshold a-wave in CSNB (Fig. 1B) (102 ± 22.52
μV) is also within normal limits (110.50 ± 10.80 μV),
suggesting that the photoreceptors (cones) are func-
tioning normally. The morphology of the supra-
threshold (Fig. 1B) ERG recorded from CSNB pa-
tients is, nonetheless, abnormal. For a high intensity
of stimulation, small oscillations (arrows 2 and 3) are
normally seen on the ascending phase of the b-wave.

These oscillations are absent in the ERG recorded
from CSNB patients giving to the ERG a “square wave” appearance. They are also minimally visible in
the recording from the father of two CSNB pa-
tients.

The scotopic ERG (Fig. 1C) in CSNB is one of low
voltage. The amplitude barely reaches 10% of that
measured in normals. The peak time of the scotopic
b-wave in CSNB appears to be faster than normal.
The results illustrated in Fig. 1D indicate that the
amount of time spent in dark adaptation influences
both the peak time and the amplitude of the scotopic
ERG recorded from normal subjects: the peak time
being faster in the early phase of dark adaptation (that
is when the rod contribution to the ERG signal is
minimal). The faster dark-adapted b-wave peak time
measured in CSNB patients might be an indication
that this scotopic ERG is mainly evoked by activating
dark-adapted cones.

The first derivative d voltage/d time of the ERG
wave is obtained by computing the voltage variation
in absolute values per unit of time for the entire du-
ration of the signal. The data found in normals and
CSNB patients are compared in Figure 2. In normals
four major components (numbered 3, 4, 5, and 6)
are identified. They represent regions where impor-
tant changes in the slope of the ERG wave occur and
correspond to arrows 3, 4, 5, and 6 of the ERGs
illustrated in Figure 1B. The most important slope change seen in the normal ERG is identified as 4; it peaks at 32 msec after flash onset (time 0) and corresponds to the peak of the b-wave of the electroretinogram. The magnitude of this slope change as a mean value of 52 μV/msec (range between 32 μV/msec-70 μV/msec). In the ERGs recorded from CSNB patients, this component has a mean magnitude of 42 μV/msec (28 μV/msec-58 μV/msec), which is within the limits of normals. On the other hand there is a complete absence of component 3 and a reduction in the magnitude of components 5 and 6 in the tracings obtained from CSNB patients. Apart from corroborating the earlier findings (Fig. 1B), the results suggest that the neuronal structure(s) responsible for the genesis of event 4 appear(s) to be functioning normally. This is verified by the magnitude of the slope change impinged and by comparing the contribution (% t) of that particular event to the building of the ERG signal. In a normal ERG event 4 contributes for 15% of the total ERG signal. In normal the same event now contributes more than 22%. This discrepancy further accentuates the finding that the suprathreshold photopic ERG in CSNB is mainly formed of that particular event. Although events 5 and 6 in CSNB are decreased in their magnitude, their contribution to the building of the ERG signal is within normal limits.

The first derivative analysis of the electroretinogram suggests that the ascending portion of the suprathreshold photopic ERG is formed of a succession of rapid slope changes, each slope change contributing to the building of the b-wave. These slope changes correspond to small oscillations on the ERG (Fig. 1B) and are identified as oscillatory potentials. These oscillations can be more selectively recorded by using a higher low-frequency cut off (100 cps instead of 1 cps). A higher low-frequency cut off will minimize the contribution of slower components of the ERG while faster ones will be accentuated. Recordings obtained using the 100 cps low-frequency cut off are illustrated in Figures 3-6.

A normal, suprathreshold, photopic OP recording (Fig. 3, tracing 1: NORMAL) is made of seven oscillations identified as N, 1, 2, 3, 4, 5, 6. They correspond to specific electrical accidents seen on the electroretinograms (Fig. 1B, NORMAL). Hence, OP2, 3, 4, 5, 6 corresponds to arrows 2, 3, 4, 5 and 6 of Fig. 1B. OPN corresponds to the leading edge of the descending a-wave, while OP1 corresponds to a slight change in the slope of the a-wave normally seen between the response onset and OP2 (tracing 2 and 3; Fig. 1B NORMALS). By comparing the normal tracings in Figure 1B with tracing 1 in Figure 3 (NORMAL), one notices that the peak of the b-wave correspond
there is a complete absence of OP2 and 3. The peak time and the amplitude of OPN, 4, 5, 6 are, however, within normal limits. The absence of OP2 makes it difficult to identify OP1 in this tracing, but the results illustrated in Figure 4 indicate that it should be present. A stimulus intensity four times above threshold (Fig. 3, tracing 3 CSNB) evokes a small wave with a peak time within the limits of OP2. This wave was identified as ?OP2 since it disappears rather than augmenting in amplitude and shortening its peak time with a graded increase in the stimulus strength. As in normal, OP5 and 6 see their respective peak time augment with an increase in the stimulus strength. There is also an augmentation in the peak time of OP4 better seen by comparing the tracings obtained at 8 and 16 times the threshold.

The separation of the complex OP (3–4) can also be achieved by lowering the intensity of the adapting

Fig. 3. Influence of a graded increase in the stimulus intensity on the light-adapted OP recorded from a normal subject (top) and a patient affected with CSNB and myopia (bottom). The intensity of the stimulus is given as a multiple of the threshold (T) and is indicated on the right-hand side of each tracing. N, 1, 2, 3, 4, 5, and 6 identify the oscillatory potentials considered in the present study. F marks the flash onset. The flash photo-artifact is seen between flash onset and OPN. Calibration bar: horizontal in milliseconds, vertical in microvolts. Bandwidth: 100–1000 cps.

corresponds to an OP made from the fusion of OP3 and OP4 into one OP, which we will identify as complex OP (3–4). Augmenting the intensity of the stimulus from 4 to 16T will separate OP (3–4) into two distinct OP. Probably as a result of this adding of one oscillation, OPN, 1 and 2 are the only signals that see their peak time shortened with an increase in intensity of the stimulus, while OP5 and 6 see theirs augmented. It is difficult to make a statement on OP3, 4 since their separation appears at an intensity value (8T) too near the maximal intensity available.

In the suprathreshold photopic OP recording obtained from CSNB patients (Fig. 3, tracing 1 CSNB)

Fig. 4. Influence of an attenuation in the intensity of the background on the OP recorded from a normal (top) and a CSNB patient (bottom). Background attenuation is indicated at the right-hand side of each tracing. The intensity of the flash is indicated at the beginning of each tracing. F indicates flash onset. The background intensity was changed from 50 lumen/m² (0.0 attenuation) to 5 lumen/m² (–1.0 log-unit attenuation). Calibration bar: horizontal in milliseconds, vertical in microvolts. Bandwidth: 100–1000 cps.
background light as illustrated in Figure 4. The top half of the figure illustrates OP recordings obtained from a normal subject. Again increasing the stimulus intensity from 2T (tracing 2) to 16T (tracing 1) while the background light intensity is kept constant and in the photopic range, will produce the splitting of the complex OP (3-4) into two distinct OP (OP3 and OP4). Dimming the intensity of the background light and keeping the intensity of the stimulus at 2T (tracing 3) will also produce the splitting of the complex OP (3-4) into OP3 and OP4. Thus a decrease in the background intensity of 1 log unit when the stimulus intensity is kept constant and slightly above threshold mimics an increase of about 1 log-unit (2T to 16T) in the stimulus intensity with the background intensity kept constant and in the photopic range. However, there are major differences. While the peak time of OP4, 5, and 6 obtained at intensity 2T, —1.0 log unit (tracing 3) falls within those measured in the tracing obtained at intensity 16T, 0.0 log unit attenuation (tracing 1); no changes in the peak time of OPN, 1 and 2 are noticed. The peak time of OPN, 1 and 2 measured at 2T, —1.0 log unit of background attenuation (tracing 3) are within the limits of those measured at intensity 2T, 0.0 log-unit of background attenuation (tracing 2) and consequently longer than those measured at intensity 16T, 0.0 log unit attenuation (tracing 1). Finally, the peak time of OP3 measured at intensity 2T, —1.0 log unit of background attenuation (tracing 3) is slightly longer than that measured at intensity 16T, 0.0 log-unit attenuation (tracing 1).

In CSNB subjects dimming of the background intensity by 1 log unit while the intensity of the stimulus is kept constant and slightly above threshold (2T), gives the same results as in normal in that it mimics an increase of about 1 log unit (2T to 16T) in the intensity of the stimulus with the background intensity kept constant and in the photopic range. A decrease in the background light intensity thus fails to produce the splitting of the complex OP (3-4) into OP3 and OP4. The peak times of OP4, 5, and 6 measured at intensity 2T, —1.0 log unit of background attenuation (tracing 3) correspond to those measured at intensity 16T, 0.0 log unit of background attenuation (tracing 1). Arrowheads in tracings 2 and 3 (Fig. 4, CSNB) point to what could be remnants of OP2 and 3. These two waves never reach the amplitude seen on the normal recordings, nor do they reach the ratio value achieved in normals when the amplitude of OP4 is compared to that of OP2 or OP3.

CSNB subjects demonstrate a loss of OP2 and OP3 in their suprathreshold photopic OP recordings. What retinal events are responsible for the genesis of these OP? The previous results suggest that OP2 is linked closely to the intensity of the flash and that regardless of the background intensity (Fig. 4). Comparing the light-adapted and the dark-adapted (after 16 min) high intensity response gives us more information (Fig. 5). Again the peak time of OP2 is not changed by dark-adapting the retina. Modification in the amplitude of OP2 is rendered very difficult to appreciate due to the tremendous increase in the amplitude of OPN. There is a complete lack in the dark-adapted signal of OP6. The peak time of OP5 is slightly augmented with DA, but no significant difference is no-
ized in its amplitude. There is a splitting of OP3 into OP3a and OP3b and of OP4 into OP4a and OP4b. This splitting is more evident in OP4. The major modification caused by dark adaptation is to produce a change in the "peak" of the response. Usually in normal the "peak" of the suprathreshold photopic OP recording is achieved with OP4. In extreme cases the amplitude of OP4 is equal to that of OP3 (measured from the baseline). In the dark-adapted retina the "peak" is achieved at the level of the complex formed by OP3a–3b.

A flicker of 30 cps is said to isolate in a preferential way the photopic or cone function.5 At least two physiologic parameters are interacting in a flicker stimulation. The first one being the rate of presentation of the stimulus and the second one the amount of light adaptation produced by each individual stimulus. Habituation of the retinal elements needs to be achieved in order to truly identify the changes produced by a flicker stimulus. This habituation is usually monitored by an absence of noticeable modifications in the recordings obtained from two consecutive flashes comprised inside a train of flashes. Averages of the sixth to the ninth flash of a flicker stimulation of increasing frequency recorded from a normal subject are illustrated in Figure 6. Increasing the rate of presentation of the stimulus from 1 cps (tracing 1) to 15 cps (tracing 4) causes profound modifications in the amplitude as well as in the peak time of OP2 and OP3. Amplitude-wise OP3 appears to be the most affected. There is no significant change in the peak times of OP4, 5, and 6. There may be, however, a slight decrease in the amplitude of the ascending portion of OP4 but not as dramatic as those observed for OP2 and OP3. The increase in the stimulus frequency appears to influence selectively OP2 and OP3 resulting in a recording that approaches that obtained from CSNB subjects (Fig. 3, tracing 1 CSNB) where a nonflickering stimulus is used.

Discussion

As it was previously reported6,7 our results indicate that the amplitude of the suprathreshold photopic ERG in CNSB with myopia is decreased when measured from the trough of the a-wave to the peak of the b-wave. However, the implicit time of the b-wave remains within normal limits. The amplitude of the a-wave, when measured from baseline to trough is not significantly different from normal. Furthermore, there is a complete absence of the two oscillations (OP2, OP3) normally seen on the ascending portion of the b-wave (Fig. 1B). This combination of a normal amplitude a-wave, normal b-wave peak time added to the absence of the two oscillations yields to an ERG wave with a rather unique morphologic presentation described by Heckenlively8 as "square-wave" like. This is probably how one could describe the ERG of CSNB patients illustrated in Figure 1B. Hill et al7 have also reported the loss of one oscillatory potential (probably OP2 in our nomenclature) in their study on CSNB. Sandberg et al13 reported a loss of the same two oscillatory potentials in the dark-adapted cone ERG of subjects affected with different forms of retinitis pigmentosa. However, in their study this loss in oscillations was always accompanied by a delay in the implicit time of the photopic suprathreshold ERG b-wave.

As expected CSNB patients demonstrated a minimal response in the dark-adapted state (Fig. 1C). Although the stimulus intensity used was in the scotopic sensitivity range, the implicit time of the b-wave ERG recorded was faster than normal. The implicit time of the b-wave ERG obtained from CSNB subjects after 16 min of dark adaptation fell within the limits of that measured in a normal subject within the first minute of dark adaptation, that is when the rhodopsin regeneration is minimal. One wonders if this observation is an indication that the b-wave ERG recorded from CSNB subjects, to a low intensity stimulus (scotopic range), after 16 min of dark adaptation, could be cone mediated. Nonetheless, this severe decrease in the amplitude of the b-wave recorded after dark adaptation, represents the physiologic measurement of what one would expect from a patient complaining of night-blindness. That is elevation of the rod threshold or, as in our case, near absence of a rod response. What is surprising, however, are the anomalies found in the suprathreshold photopic ERG of CSNB patients. It appears from the first derivative analysis (Fig. 2) as well as from the selective recordings of the oscillatory potentials (Fig. 3) that apart from the absent OP2 and OP3 in the ERG of CSNB patients, the other ERG subcomponents (OPN, 1, 4, 5, 6) are within the normal limits both in amplitude and in peak time. What are the retinal mechanism(s) responsible for the genesis of these two OP? Since these anomalies are seen in the photopic ERG of CSNB subjects, does it imply that their photopic system is also deficient? Or does it suggest that the retinal mechanisms, normally involved in night vision, those that are defective in CSNB with myopia could also be, in a normal retina, implicated in the genesis of the suprathreshold photopic ERG? Some indications are provided in the results illustrated in Figures 3–6.

OP2

The peak time of OP2 will shorten with an increase in the stimulus strength (Fig. 3). An increase in the intensity of the stimulus will also produce an increase in the amplitude of OP2. However, dimming the in-
tensity of the background light (Fig. 4) will not modify either the peak time nor the amplitude of OP2, while OP4, 5 and 6 will behave as if there has been an increase in the intensity of the stimulus. Along those lines dark adaptation (Fig. 5) has no influence on the peak time of OP2 and probably no effect on its amplitude. Finally, OP2 is one of the two OP that demonstrate significant modification to a slow flicker (Fig. 6). Thus, as long as the intensity of the stimulus is kept constant, the level of light adaptation appears to have very little, if any, influence on the genesis of OP2. Hence, OP2 appears to be a very good indicator of the intensity of the stimulus and that regardless of the level of light adaptation the retina is subjected to (signal to noise). This is in direct opposition with the behavior of OPN where dimming the intensity of the background (Fig. 3, tracing 2–3, normal) or dark adaptation (Fig. 5) will cause a significant increase in its amplitude. Correspondence between ERG waves (Fig. 1B) and OP recordings (Fig. 3) indicates that OPN would signal the first portion (leading edge) of the negative a-wave. Since the a-wave is said to reflect the activation of the photoreceptors,14,15 OPN would be the oscillatory potential counterpart signalling this activation. If the ERG when taken as a succession of different electrical events (Fig. 2) or the seven oscillatory potentials that correspond to them (Fig. 3) do reflect in a chronologic order the sequence of activation of more distal (from the photoreceptors) retinal elements, as suggested from microelectrode study,16 then the generator(s) of OP2 would be located after the photoreceptors but before the generator(s) responsible for the peak of the suprathreshold photopic b-wave. Finally, in view of the above mentioned findings the neural structure(s) responsible for the genesis of OP2 would be involved in a retinal pathway responsible for keeping constant the signal to noise ratio.

OP3

This oscillatory potential is fused with OP4 at low but still higher than threshold stimulus intensity (Fig. 3). Increasing the stimulus intensity or decreasing the intensity of the background light (Fig. 4) will provoke the splitting of complex OP (3–4) into two distinct OP, namely OP3 and OP4. OP3 is, of the seven photopically evoked oscillatory potentials, the one that is the most affected by a slow flicker (Fig. 6). Both its peak time and its amplitude are modified severely. Finally, dark-adaptation (Fig. 5) will augment its relative contribution to the overall signal obtained under such conditions, making it the major OP in the dark-adapted signal (while OP4 is the major one in the light-adapted response). It is well known3 that a flicker stimulation of a frequency rate of about 30 cps will isolate the cone system in a preferential way. This notion is based on the fact that rods are incapable to follow repetitive stimuli at a rate exceeding 10 cps. Thus, the flicker-induced modifications of OP2, and to a greater extent of OP3, added to the potentiating effect that dark-adaptation exert on OP3 suggest that the genesis of OP3 could be in very close relation with the scotopic (rod) function. If this is the case then OP3 normally recorded using a suprathreshold stimulus delivered under light adaptation would be rod dependent. This hypothesis would explain why OP3 is missing in the recordings obtained from CSNB subjects.

If, as it is suggested from our results, there is a rod contribution to the photopic ERG, is it due to the fact that the intensity of the background used was not bright enough to bleach entirely the rods? Normally, the reported2,5–6 high intensity white light photopic b-wave amplitude is in the range of 150–200 µV, with a peak time of about 26 msec. Ours ranged between 180–260 µV, with a peak time of about 32 msec. It is well known5,17 that an increase in the stimulus intensity will produce an increase in the amplitude of the b-wave but as it is demonstrated in Figure 3 an increase in the intensity of the stimulus will also produce an increase in the implicit time of the oscillatory potential responsible for the peak of the photopic b-wave. This rather unusual behavior is only seen in the light-adapted ERG and has already been examined by previous investigators.17 Thus, our higher amplitude and longer peak time reported (when compared with other studies) would find some explanation in this unique behavior. Furthermore, the morphology of the suprathreshold photopic ERG wave recorded in normals (Fig. 1B) is comparable to those illustrated in other studies.18 And it is to an anomaly in the morphology of the suprathreshold photopic ERG wave recorded from patients affected with CSNB and myopia that the present report is addressed to.

In conclusion, a decrease in the amplitude of the ERG while the implicit time of the b-wave remains normal is usually interpreted as reflecting a focal destruction of retinal elements.3,17,19 For instance a low voltage photopic ERG with a normal b-wave peak time would be interpreted as a partial destruction of the cone population. The normal peak time suggests that the remaining cones are functioning normally. However, it is important to remember that the Ganzfeld-evoked ERG is a mass response resulting in the homogeneous activation of a large area of the retina. In that respect we believe that destruction of a portion of the retinal tissue or a decrease in the number of photoreceptors being evaluated should be reflected on the ERG wave by an equal attenuation of all the electrical segments that compose the ERG wave. This
equal reduction is probably seen in sector retinitis pigmentosa19 or chorioretinal scar,17 although no attempt was made to analyze the wave recorded. The photopic suprathreshold ERG recorded from patients affected with CSNB and myopia is decreased in amplitude with a normal b-wave implicit time. Analysis of the ERG subcomponents (Fig. 2, 3) revealed that the two oscillations (OP2 and OP3) normally seen on the ascending portion of the b-wave were absent from CSNB recordings. However, the remaining ERG subcomponents (OPN, 1, 4, 5, and 6) appeared to be within normal limits both in amplitude and in peak time (Figs. 2, 3). The first one (OP2) would transduce the electrical activation of retinal element(s) involved in keeping constant the signal to noise ratio by truly monitoring the real intensity of the stimulus regardless of the state of light adaptation the retina is in. The second one (OP3) appears to be in close relation with rod activity. Both generators would be located between the photoreceptors and the bipolar-Müller cells layer. No indications are provided, however, on how this deficit is achieved in CSNB. Among the possibilities there are: lack in the retinal structures responsible for the genesis of OP2 and OP3, neurotransmission (or neurotransmitter) deficiency, or a strong inhibitory mechanism which would be rendered more potent in CSNB. Finally, it was suggested by the work of Sandberg et al20 that a rod-cone interaction was possible in the distal (from the ganglion cells) human retina, namely that the rod function could influence the cone function and that this interaction would be reflected in the temporal aspect of the cone b-wave. One wonders if OP2 and/or OP3 could not, in part, be the result of such an interaction and, consequently, they would be generated only if such an interaction is possible, that is only if the two retinal channels (scotopic and photopic) are functional.

Key words: photopic, electroretinogram, oscillatory potentials, night blindness, rod-cone interaction, retina

Acknowledgments

Special thanks to all the CSNB patients for their kind cooperation, to Dr. S. Molotchikoff for his helpful comments, and to Suzanne Dobby for her patient typing of the manuscript.

References