Original Article

Electrophysiological Findings in Persons With Nyctalopia

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Abstract

Introduction: Ophthalmologists are occasionally confronted with an individual presenting with nyctalopia (i.e., a relatively greater difficulty seeing at night). When there is no accompanying abnormality seen in the fundus, visual electrophysiology becomes useful as an objective means of assessing rod (scotopic) photoreceptor function or pathway defects. Materials and Methods: A retrospective study was performed on 50 consecutive patients, aged less than 40 years, with seemingly normal fundi and good vision [visual acuity (VA) >6/12] presenting to the Visual Electrodiagnostic Unit, Singapore National Eye Centre, for the investigation of nyctalopia over a 2-year period. Subjective scotopic threshold sensitivity (STS) and objective full-field electroretinogram (ERG) were performed. Persons with abnormal test results were identified. Results: Normal ERG scotopic responses were obtained in 74% of subjects. There was no significant difference in age, refraction and STS levels between subjects with abnormal and normal ERG. In the group with abnormal scotopic ERG responses, 9 were identified to have non-specific rod dysfunction, 2 had rod-cone dystrophies and 2 had ERG changes suggestive of congenital stationary night blindness (CSNB). Conclusion: A large number of subjects presenting with nyctalopia had normal ERG findings. We can only assume that in these patients, no significant rod pathway dysfunction exists and that optical (e.g., night or instrument myopia) and psychological aetiologies should be considered. The fact that an abnormal result occurs in 26%, however, suggests that ncytalopia should be evaluated with electrophysiolgoical testing even when the fundi appear normal.

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Introduction

Nyctalopia is a subjective sensation of poor vision under dark conditions. It may be clinically associated with retinal diseases affecting the rod photoreceptors or post-receptoral pathways. Retinal changes are often visible in the presence of a photoreceptor dystrophy. However in some cases, changes can be subtle and the fundi may even appear entirely normal. When the condition is longstanding or gradual in onset, the subject may report very few symptoms. A problem may be recognised only when visual difficulties are encountered under specific conditions. In Singapore, it is most commonly noted in young men serving National Service, when they have problems during night shooting or night exercises, and in adults when driving at night, or in dimly lit underground carparks.

In this study, we reviewed all patients who presented to the Visual Electrodiagnostic Unit, Singapore National Eye Centre with symptoms of nyctalopia but absence of obvious retinal (fundal) pathology. The aim of this study was to determine how often electrophysiological evidence of retinal pathology was present in this group of patients.

Materials and Methods

A retrospective review was made of all patients who presented to the Visual Electrodiagnostic Unit for the investigation of nyctalopia during the years 2003 to 2004. Only those patients aged less than 40 years, with best corrected visual acuity of 6/12 or better, and who were found to have normal looking fundi were included in this study.

Factors such as age, gender, refractive error and visual acuities were noted. Refractive error was taken as that recorded in their prescription for spectacles. If no spectacles were used, the refractive error was recorded as being plano (0D). Scotopic threshold sensitivity, and full-field electroretinogram (ERG) studies were obtained.

The scotopic sensitivity test (SST) is a measurement of final dark adapted threshold. It was tested using the SST-1 (LKC Technologies Inc, MD, USA). A mini-Ganzfield bowl to present a full-field, slowly oscillating (0.5 Hz),

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sinusoidal white light of variable intensity to the patient. SST was not routinely done on all patients. Patients were selected to perform SST when time permitted. Subjects were dark adapted for at least 20 minutes in a completely darkened room. To obtain accurate threshold and maintain the level of consistency, the light was presented to the patient using a 2-alternative procedure. In this procedure, the patient was presented with 2 trials, one of which included a stimulus and the other a blank (no stimulus). The patient was asked to make a forced choice (i.e., asked which trial contained the stimulus), or to indicate if they could not tell. The light was first presented at 0dB. If the light was not seen, the light intensity was increased in 5dB increments until the stimulus had been made sufficiently bright that the patient could see the light. The light intensity was then reduced in 1dB steps until the patient could no longer see the light. The resulting value was used for the patient's scotopic sensitivity. A threshold of more than 5dB was considered abnormal.

The full-field ERG was recorded using the Espion system (Diagnosys, LLC, Littleton, MA, USA) with Dawson-Trick-Litzkow (DLT) electrodes. Pupils were dilated with tropicamide 1% and the test was commenced when both pupils were equally dilated to at least 7 mm. The testing protocol strictly adhered to International Society of Clinical Electro-physiology of Vision (ISCEV) standards and consisted of 5 responses, namely, the scotopic, maximal, oscillatory potential (OP), photopic and 30 Hz flicker.[1] The scotopic, maximal and OP responses were obtained after dark adaptation for at least 20 minutes. The photopic and 30 Hz flicker responses were recorded after 10 minutes of light adaptation. The response parameters (amplitude and implicit time) of each individual were then compared with the laboratory's normative data to screen for photoreceptor dysfunction or transmission defect. Scotopic b-wave amplitudes of >130uV were assumed to be within normal limits.

Subjects were divided into 2 groups of normal and abnormal ERG findings. The difference in age, refraction and STS levels were analysed using independent *t*-test and the difference in the proportion of genders using Chi-square test. Correlation between STS and scotopic b-wave amplitude was analysed.

Results

Of the 53 patients who presented for the investigation of nyctalopia, 51 satisfied the inclusion criteria of this study. Investigation was not possible in one subject because of the subject's inability to tolerate the placement of electrodes. Of the remaining 50 subjects, 48 were male (96%) and 2 female. The mean ± SD age was 22.5 ± 4.9 years. Thirty-seven (74%) patients had an ERG response that was well within normal limits (Table 1). There was no significant difference in age of presentation, gender, refractive error or scotopic threshold sensitivity recordings in the groups with normal or abnormal ERGs.

Of the 13 patients who returned an abnormal response, 9 patients had rod responses which were below the normal limits but not delayed (i.e., non-specific rod dysfunction). Two patients had concurrent delayed implicit times suggestive of a rod-cone dystrophy. One patient had an ERG response suggestive of the complete form of congenital stationary night blindness (CSNB), and another of incomplete CSNB (Fig. 1).

There was no significant difference in refractive error between groups, although the range of myopia was greater in the "normal" group (Table 1, Fig. 2).

The subjectively measured scotopic threshold sensitivities (STSs) were available in 35 subjects. A range of normal and abnormal STSs were present in each outcome group. Abnormal STS was noted in 40.7% (11/27) of those with normal ERG findings, and 75% (6/8) with abnormal ERG result. Scotopic b-wave response was poorly correlated to STS (Fig. 3, R2 = 0.035). One subject with scotopic b-wave amplitude well within the normal limit had a STS of 26dB; whilst one with no recordable ERG response had a STS which was normal.

Discussion

Persons with nyctalopia or night blindness present with a relatively greater inability to see in dim or dark conditions. The visual system in humans is split into one that sees in

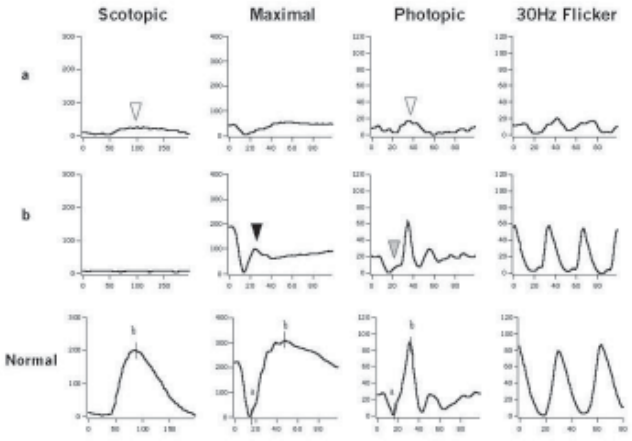

Fig. 1. Abnormal ERG responses.
a. Rod-cone dystrophy. Note reduced and delayed a- and b-wave responses under both scotopic and photopic conditions (open arrows).
b. Congenital stationary night blindness. Note absent scotopic response, and electronegative maximal response (dark arrow) where the b-wave is reduced below baseline. The photopic a-wave is also broadened (grey arrow) secondary to abnormality of the on-response.

Table 1. Characteristics of Patients Within Each Outcome Group

	Normal	Abnormal	*P*
No. (%)	37 (74%)	13 (26%)	
Age (y)	22.8 ± 5.3	22.0 ± 3.6	0.6279
Gender (M:F)	35:2	13:0	0.3922
Refraction (D)	-4.3 ± 3.6	-3.5 ± 3.2	0.5256
Scotopic threshold sensitivity (dB)*	7.7 ± 4.7	8.2 ± 3.8	0.7865
Scotopic b-wave amplitude (uV)	180.4 ± 61.0	97.4 ± 67.6	<0.001

* Scotopic threshold sensitivity was available in only 35 subjects

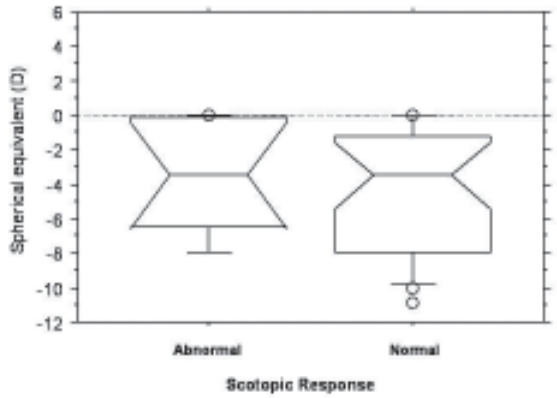

Fig. 2. Refractive error in different outcome groups.
Central line denotes mean with box marking the limits of the 25th and 75th percentiles. The lines denote the limits of the 10th and 90th percentiles with the circles representing outliers beyond the 10th and 90th percentiles.

bright conditions (photopic) and another that functions in the dark (scotopic). The photopic system consists of cone pathways, and is responsible for coloured, high-definition vision. In contrast, the scotopic system consists of rod pathways, and results in the grey-scale, low-resolution vision we have under dark conditions.

When a person presents with nyctalopia, the principal concern is to exclude any retinal pathology. The scotopic ERG serves just this purpose, and is a useful and objective way of assessing rod pathway function. Each laboratory has its own age-specific normal limits, unique to its ethnic population and equipment. Normal data are obtained by collecting responses from a volunteer population of normal subjects. The normal limits were defined as that 2 standard deviations about the mean (i.e., with a confidence intervals of 95%).

In this study, some 74% of subjects presenting with nyctalopia had scotopic ERG within normal limits. There was poor correlation between the amount of scotopic response loss and the subjective degree of nyctalopia (Fig. 3). The degree of nyctalopia can be measured using the STS. However, being a subjective measure, the STS is

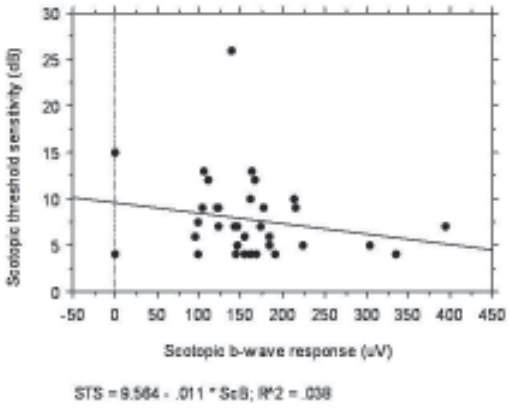

Fig. 3. Scotopic threshold sensitivity in different outcome groups.
Normal values: STS <5dB, Scotopic b-wave amplitude >130uV

notoriously unreliable. Indeed, an abnormal STS was noted in 59% of subjects with normal scotopic ERGs, and a normal STS response was noted in 12% of subjects with abnormal ERGs.

Why would persons with apparently normal scotopic responses complain of nyctalopia? Several explanations may exist. "Night myopia", for example, is a very well recognised phenomenon,[2] and occurs as a result of over-accommodation and pupil dilation under dark conditions. This is an optical problem and typically, objects may become more blurred, with halos noted about lights, and ghosting around objects. Pre-existing myopia may compound the problem.[3,4] Sixty-eight per cent of all subjects and 70.2% of "normal" subjects had some degree of myopia.

Many of these patients are young men who notice a problem when they begin their national service and are required to participate in night manoeuvres or target-shooting. This accounts for the high proportion of males in this study. To aggravate matters further, when these people are required to look down gun-sights, binoculars or scopes, "instrument myopia" may occur.

Psychological problems may also exist. With the widespread availability of electrical lighting, present generations may be more accustomed to working in well-lit environments and unaccustomed to functioning in the dark, where vision is naturally poorer. Stress and fatigue may also play a factor. There may also be a functional element with some subjects claiming to have nyctalopia so as to avoid certain aspects of their training.

Nyctalopia is easier to explain when some scotopic abnormality is noted. However, the significance of a mildly decreased scotopic response not associated with increases in implicit times can be difficult to interpret. Since ranges are computed using 2-sided 95% confidence intervals, 2.5% of persons with ERG results below the normal limit

may actually be normal. Furthermore, the ERG response is critically dependent on the amount of light entering the eye. Small variations in pupil size, eye closure or blink artifact can significantly affect the level of response obtained. During the test, individuals must be closely watched. In uncooperative subjects, no reliable conclusion can be drawn. Lower ERG responses are also known to occur in older patients, and in those with high myopia. All these factors need to be taken into consideration during the analysis of the recorded response so as not to return a false positive result.

The diagnosis is simpler when there is significantly decreased scotopic amplitude or delay in implicit time, the combination of which suggests a rod dystrophy (Fig. 1a). In some cases, the cone (photopic) system may also be involved either at presentation or over time. In early cases, the fundal changes commonly seen in rod-cone dystrophies may not be apparent. However, subtle perivascular pigmentary changes, optic disc pallor and vessel attenuation may be seen.[5]

Finally, 2 subjects had responses suggestive of CSNB. Two forms exist (complete versus incomplete), each with typical ERG appearances (Fig. 1b).[6,7] Fundal appearance is typically unremarkable. The complete form has a typically absent scotopic ERG, an electronegative maximal ERG, broaden photopic a-wave and an abnormal on-response. The incomplete form has a diminished scotopic ERG, and both on- and off-response abnormalities.

When confronted with an individual with nyctalopia who has an otherwise normal ocular examination, we need to assess all aspects of the problem. This involves determining the exact nature of the visual complaint. Important features may include:

1. The nature, duration and severity of the problem.
   Patients with rod dysfunction may complain of darkness, or of requiring a longer time to adapt to dark conditions than their peers. Optical problems (like night myopia or aberrations) are more likely to cause blurring, halos or ghosting.
2. Other visual symptoms which may be related to a photoreceptor abnormality.
   Cone dystrophies may be associated with the loss of visual acuity, photopsias and colour deficits. Rod dysfunction may be associated with constricted visual fields.
3. Exact situations when problems are encountered.
4. Presence of family history.
5. Detailed medical and medication history.

Seventy-four per cent of our subjects with "nyctalopia" but seemingly normal fundi returned normal scotopic responses. ERG remains the best available test to objectively assess retinal function. Reliable testing is occasionally limited by patient co-operation, and a repeat ERG may be necessary. However, till there is some alternative method of accurately measuring the subjective levels of nyctalopia, we can only assume that if scotopic ERG is within normal limits, no significant rod pathway dysfunction exists. The possibility of optical or psychological aetiologies should be considered. The fact that an abnormal result occurs in 26%, however, suggests that ncytalopia should be evaluated with electrophysiolgoical testing even when the fundi appears normal.

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