Principles and Practice of Clinical Electrophysiology of Vision

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PART XII

Principles of Clinical Testing

61

Testing Levels of the Visual System

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Noninvasive clinical electrophysiological and psychophysical measurements allow an assessment of the health of almost the entire length of the visual system. An understanding of each test and their interrelationships assists the diagnosis of a number of diseases.[19] This is facilitated by the layered nature of the visual system and the assignment of electrical potentials from specific tests to particular cell layers. Specific tests can now be done to assess the retinal pigment epithelium (RPE) through the retina to the visual cortex.

Because most evoked responses stimulate the entire retina, regional loss of function may not be noticeable in the test results. By adjusting stimulus conditions and techniques of recording, a representation of the sequence of events along the visual pathway can be recognized and the cone and rod systems measured separately. If the stimulus has an abrupt onset (as is usual), the delay between stimulus and response indicates additional signs of dysfunction that are not elicitable by other methods (e.g., psychophysics).

An understanding of the neuronal response order (from photoreceptor to visual cortex) and the interrelationships of the responses is important since an abnormality at a lower level will usually give an abnormal response further along the sequence chain, and misleading interpretations can be made if abnormal test results are inappropriately taken out of context. For instance, an abnormal visual evoked cortical potential (VECP) might be found in a number of retinal dystrophies and could be misinterpreted if an electroretinogram (ERG) is not performed. *Therefore, after taking an initial history from the patient, the tests to be performed on the patient should be ordered in such a manner as to maximize the success in diagnosing the level of dysfunction.* For instance, if the patient has an abnormal ERG, then a VECP is seldom necessary. In practice this approach must also be modified to maximize patient flow; in some clinics, tests are scheduled on different days, and patients who require further tests are recalled. In other cases, a VECP will be done before the ERG because the latter requires pupillary dilatation and the patient cannot be recalled.

While the retina is composed of a complex neuronal matrix with both vertical and horizontal components, it is possible to detect abnormalities of the rods and cones, the photoreceptor layer generally, the middle retina, and the ganglion cell layer (Table 61–1). If the central retina is reasonably healthy, then the VECP can detect abnormalities in the conduction of retina-generated signals to the visual cortex.

A directed approach to visual system evaluation is

TABLE 61–1.
Localization of Lesions by Electrophysiological Testing*

Location	Test
Retinal pigment epithelium	Electro-oculogram
	DC ERG c-wave
	C-wave of the ERG (if the amplifier bandwidth is sufficient)
Outer segments	Early receptor potential
	Densitometry
Receptor layer	ERG a-wave (in general)
Cone system	Photopic ERG
	Color vision testing
	Flicker ERG
Rod system	Rod-isolated ERG
	Dim blue stimulus or white stimulus below the cone threshold
	Dark adaptation testing
Middle retinal layers/Müller cells	ERG b-wave
Amacrine/bipolar cells	Oscillatory potentials
	Pattern ERG (P50)
	Threshold negative response
Ganglion cell layer	Pattern ERG
Macula†	Focal ERG (specialized test)
Optic tract‡	Visual evoked cortical potential

*Details of each test can be found in the respective chapters.
†The flash ERG results in a panretinal response, and the macula only contributes 10% to 15% to the b-wave amplitude.[17] If a patient with a macular lesion has a poor photopic response, then the patient has a problem affecting the entire cone system.
‡If visual acuity is reduced, the VECP is reduced in amplitude and delayed no matter what the cause such as refractive errors or retinal disease; thus an abnormal VECP under these circumstances can be confused with a neuropathy unless a clinical examination is made.

especially important for disorders that either have specific sites that have been shown to be abnormal on histopathology of the suspected disease or are believed to be abnormal on the basis of clinical appearance or other tests. For example, the ERG is well suited to evaluate generalized photoreceptor disease such as retinitis pigmentosa (RP) because the response is a mass one that is initiated by photoreceptors. Widespread photoreceptor dysfunction and loss are early features of RP. Best's macular dystrophy, on the other hand, has presumably a diffusely abnormal RPE, with inclusions of lipofuscin on histopathology but normal photoreceptors (except in regions of RPE loss or scarring). Thus, it is not surprising that the ERG is normal in Best's dystrophy but the electro-oculogram (EOG), which tests potentials generated by the RPE, is abnormal.

While Table 61–1 is helpful in forming a perspective on how the various tests can be used to evaluate cellular layers or "minisystems" in the visual system, it is useful, despite the risk of redundancy, to tabulate each test with the information that can be expected from doing it. These are presented in Table 61–2.

THE RETINAL PIGMENT EPITHELIUM

The two main tests for evaluating the RPE are the EOG and the c-wave of the direct-coupled electroretinogram (DC-ERG) (see Chapters 39 and 42). The EOG electrodes, placed on the inner and outer canthi of each eye, measure the standing potential of the pigment epithelium as the patient fixates regularly between two points 30 degrees apart. This standing potential normally fluctuates but decreases with dark adaptation; subsequent light adaptation causes a large slow oscillation increase. The maximum value measured during light adaptation divided by the smallest dark-adapted value represents the Arden light peak/dark trough ratio, which is widely used to assess RPE function.

In certain disorders the slow oscillations of the EOG can be abnormal to varying degrees while the ERG is invariably normal or near normal. Examples of such diseases would include Best's vitelliform macular dystrophy, Stargardt's disease or fundus flavimaculatus, dominant drusen, and pattern dystrophy of the RPE. Of interest, the fast oscillations of the EOG are preserved in Best's macular dystrophy, even at a time when the slow oscillation is markedly abnormal.[18]

In other disorders such as cone dystrophy, the EOG is normal, but the ERG is abnormal. In some forms of cone dysfunction the standing potential of the eye may be subnormal even though the light-induced slow rise of this potential is of a normal absolute value. This results in a light-to-dark ratio for the slow oscillation that is greater than normal. Supernormal EOG light-to-dark ratios have been reported in X-linked cone dystrophy and cone dysfunction secondary to digoxin retinal toxicity.[19, 21]

In certain subtypes of RP, the slow oscillations of the EOG may be relatively preserved. For example, type I RP, which is associated with early diffuse severe rod dysfunction, is usually associated with an abnormal EOG slow oscillation early in the disease.[18] On the other hand, type II RP, which is associated with regional loss of rod and cone function (often a cone-rod loss pattern) is more likely to have preservation of the EOG slow oscillation. Of importance with regard to the site of pathology, the fast

TABLE 61–2.
Tests of Visual Function, Information Obtained, and Diseases Where Tests Are Informative

Test	Location/Information	Conditions Investigated
Visually evoked cortical response (using various types of stimuli)	Integrity of the primary and secondary visual cortex	Cortical blindness Malingering Assessment of visual acuity
	Proportion of crossed and uncrossed functional fibers in the chiasm (retinocortical projections)	Albinism[1,7] Prader-Willi syndrome[2] Septo-optic dysplasia Pituitary syndromes
	Continuity of optic nerve and tract radiations	Congenital defects Inflammation, injury Other optic atrophies Toxic neuropathy
	Demyelination	Multiple sclerosis Leukodystrophies
Pattern evoked ERG	Amacrine and ganglion cell layer of the retina	Glaucoma Diabetic retinopathy Early maculopathies Traction on the macula
Components of the Flash ERG		
Oscillatory potentials	Amacrine cells, possibly horizontal, interplexiform cells	X-linked and autosomal recessive congenital stationary night blindness Defects of neurotransmission, Parkinson's disease, autism, drug toxicity
	Indicator of microvasculature status in the middle retinal layers	Diabetes mellitus Central vein occlusion
b-wave	Müller cells Bipolar cells	Disorders with negative ERG (CSNB, retinoschisis, quinine toxicity, etc.)
a-wave	Photoreceptors	Retinitis pigmentosa and other generalized retinal degenerations
c-wave	Hyperpolarization of apical membrane RPE	Diffuse RPE disease
Electro-oculogram[18]		
Fast oscillations	Hyperpolarization of basal membrane RPE after periodic light stimulation	Retinitis pigmentosa Diffuse RPE disease
Slow oscillations	Slow depolarization of the basal membrane of the RPE with light after dark adaptation	Best's macular dystrophy Stargardt's disease Dominant drusen Chloroquine toxicity Retinitis pigmentosa
Psychophysical Tests[8]		
Sustained spatial interaction (Westheimer sensitization-desensitization paradigm)	Inner and outer layer plexiform layers	Age-related macular degeneration
Transient spatial interaction (Werblin windmill paradigm)	Inner plexiform layer Dopaminergic amacrine	Parkinson's disease Effect of haloperidol on Tourette syndrome and schizophrenia
Rod-cone interactions	Rod-mediated inhibition of cone function	Some forms of CSNB Certains types of retinitis pigmentosa and related disorders
Cone-cone interactions	Cone-mediated inhibition of cone function (possible horizontal, interplexiform roles)	Currently unknown clinically
Dark adaptometry	Kinetics of dark adaptation and final sensitivity of rods and cones	Night blindness (occasionally due to cone dysfunction)

oscillations of the EOG appear abnormal earlier than do the slow oscillations of the EOG in some forms of RP.[18]

The large c-wave found with DC ERG testing correlates highly with the EOG Arden ratio when measured concurrently in patients in a number of different diseases.[16] Current techniques for measuring the c-wave are difficult to employ in the clinical setting, so the test is not widely used, even though it is a good measure of RPE function.

THE EARLY RECEPTOR POTENTIAL

The early receptor potential (ERP) is evoked by an intense stimulus flash. It occurs with no detectable latency and precedes the a-wave. The initial corneal positive R1 has been correlated with the conversion of lumirhodopsin to metarhodopsin I.[15] The following negative R2 corresponds with the conversion of metarhodopsin I to metarhodopsin II.[6] The corneal voltage results from charge displacement across the outer limb membrane due to changes in the conformation of visual pigment. Capacitative current then flows from (or to) the inner limb. Cones contribute disproportionately to the ERP since more of their pigment is present in the true surface membrane. In humans, Goldstein and Berson found that of the total ERP amplitude, 60% to 80% was generated by cones and 20% to 40% by rods.[10] In order to record the ERP, investigators have used a lens filled with saline solution coupled to the electrode in a side arm that is covered with black tape in order to eliminate the photovoltaic artifact that occurs.[4] Because there is no neural amplification, flashes that elicit measurable ERPs bleach a considerable proportion of the pigment in the retina, and interstimulus intervals should be at least several minutes. The ERP has been tested in a number of retinal dystrophies, and while delays have not been found, amplitude reduction has been observed that corresponds to a decrease in the quantity of visual pigment. Several types of RP have shown quicker regeneration time than normal.[3] The clinical useful of the ERP is limited since the test has not been shown to be diagnostic of any particular disease and can only be performed in a limited number of research centers.

ELECTRORETINOGRAM

A number of studies[5] have been performed that correlate the mass ERG response to intracellular and intercellular retinal recordings, and there is general agreement that the a-wave is generated by the receptor layer and the b-wave is generated by Müller cells as a sequela to bipolar cell activity. While there is a temptation to think simplistically about origins of the ERG responses, various studies suggest complicated interactions involving inhibitory and excitatory cells affecting the receptor–bipolar-ganglion cell junctions and cell bodies. Details can be found in Chapters 5 and 7.

ELECTRORETINOGRAPHIC WAVEFORM CHANGES CHARACTERISTIC OF DISEASE

There are several changes in waveform that, when present, greatly help in arriving at a diagnosis. These include a loss of oscillatory potentials, a negative response, i.e., a large a-wave and reduced b-wave (Table 61–3), the combination of a very abnormal photopic ERG with a normal rod ERG, and a highly abnormal cone and rod ERG. Specific loss of oscillatory potentials has been notably associated with X-linked and autosomal recessive congenital stationary night blindness, vascular ischemia, and occasional cases of cone dysfunction and retinotoxicity, e.g., to ethambutol. The negative waveform, seen under test conditions of dark adaptation when using a bright flash stimulus, is a well-formed negative a-wave and a b-wave that may reach the isoelectric point. Negative a-waves are most commonly seen in cases of juvenile retinoschisis and congenital stationary night blindness, but other disorders are in the differential diagnosis and are presented in Table 61–3. Historically the term "negative waveform" has been applied to the scotopic bright-flash ERG, but many disorders can produce such a waveform in the photopic ERG as well.

Various degrees of negativity may be found. Thus, in some cases of retinoschisis and congenital night blindness, the b-wave is entirely absent, and the ERG consists of a P_{III} only. There is considerable amplification at the photoreceptor-bipolar synapse, so small photoreceptor responses produce large b-waves, and the latter "saturate" for flashes of intensity sufficient to evoke a measureable a-wave. The isolated PIII response is thus only seen with bright flashes under scotopic conditions.

Inferences about the site of pathological lesions can be made by considering at what level the dysfunction appears to occur and by comparing various tests of different levels. Thus, in juvenile retinoschisis the patient typically has a large a-wave and a poor b-wave, which suggests that the dysfunction lies after the photoreceptor layer. Likewise, in congenital stationary night blindness, which also has a large negative wave (see Tables 61–3 and 61–4), the rod ERG varies from very small to nonrecordable, while the dark-adapted bright-flash ERG has a large a-wave and poor b-wave, again suggesting a problem in the rod system distal to the rod photoreceptor. A poor cone response (either flicker or photopic ERG) in face of a full visual field and good rod response is typically seen in cone degenerations or dystrophies.

TABLE 61–3.
Disorders With "Negative" ERG Waveforms*

	Scotopic ERG		Photopic ERG	
Disorder	a-Wave	b-Wave	a-Wave	b-Wave
Stationary defects				
Autosomal recessive CSNB†	N‡	↓ ↓ ↓	N	± ↓
X-linked recessive complete CSNB	N	↓ ↓ ↓	N	± ↓
X-linked recessive incomplete CSNB (Miyake)	N	↓ ↓	N	± ↓ ↓
Åland disease (Forsius-Eriksson ocular albinism)	N	! ↓	N	± ↓ ↓
Oguchi's disease	N	± ! ↓	N	± ↓
Retinal dystrophies				
Early or transitional forms of RP	± ↓	↓ ↓	± ↓	± ↓
Infantile Refsum's disease	↓ ↓	↓ ↓ ↓	↓ ↓	↓ ↓ ↓
Goldmann-Favre vitreoretinopathy	↓ ↓	↓ ↓ ↓	± ↓	↓ ↓
X-linked recessive retinoschisis	N–↓ ↓	± ↓–↓ ↓ ↓	± ↓	± ↓–↓ ↓
Congenital blindness				
Leber's congenital amaurosis	↓ ↓	↓ ↓ ↓	± ↓ ↓	↓ ↓ ↓
Vascular disorders				
Ischemic central vein occlusion	± ↓	± ↓ ↓	NI	NI
Central retinal artery occlusion	± ↓	± ↓ ↓	± ↓	± ↓ ↓
Retinal Toxicity				
Quinine	± ↓	↓ ↓	± ↓	± ↓ ↓
Vincristine	N	↓ ↓ ↓	N	N
Paraneoplastic melanoma	± ↓	↓ ↓ ↓	NI	NI§
Optic Atrophy	N	± ↓ ↓	N	± ↓
Degenerative myopia	± ↓	± ↓ ↓	± ↓	± ↓ ↓

*Adapted from Weleber RG, Pillers DM, Hanna CE, Magenis RE, Buist NRM: *Arch Ophthalmol* 1989; 107:1170–1179; Francois J: *Int Ophthalmol Clin* 1968; 8(4):929–947; and Black RK, Jay B, Kolb H: *Br J Ophthalmol* 1966; 50:629–641.
†CSNB = congenital stationary night blindness.
‡N signifies normal amplitudes. NI indicates no information. The "±" sign indicates amplitudes variably reduced. Down-pointing arrows indicate severity of the subnormality, with a single arrow signifying mildly to moderately decreased amplitudes, two arrows denoting markedly decreased amplitudes, and three arrows for severely decreased amplitudes.
§Thirty-hertz flicker ERG responses were normal.

A comparison of tests is often useful in localizing the level of the visual system that is primarily affected. Table 61–4 lists multiple examples of this phenomena. An abnormal EOG and a normal ERG suggest RPE dysfunction, such as in Best's disease. A normal ERG with an abnormal VECP (assuming macular function is present) strongly suggests an optic neuropathy or retrochiasmal problem. An abnormal pattern ERG in the face of a normal VECP and flash ERG suggests ganglion cell dysfunction, as might be seen in early glaucoma, and also occurs in conditions like Stargardt's disease before visual acuity is noticeably depressed.

Interpretation of the VECP and ERG may help to differentiate primary optic atrophy from optic atrophy secondary to retinal degeneration, but occasionally this comparison may be difficult in situations, such as in long-standing advanced glaucoma, where results of both tests may be abnormal. Also, some disorders such as dominantly inherited optic atrophy may have mildly abnormal ERGs as well as abnormal VECPs. Nevertheless, the combination of VECP and ERG is very useful to determine the site of pathology, especially in blond fundi in the face of subnormal visual acuity.

For a limited number of indications, the combination of VECP and pattern ERG (PERG) is extremely helpful. Disorders that produce dysfunction of ganglion cells but good acuity, such as early glaucoma, are most likely to produce abnormalities of the PERG with lesser defects or normal VECPs. This results from the fact that relatively few intact macular fibers are needed to produce an intact VECP, whereas current studies suggest that ganglion cell

TABLE 61–4.
Examples of Definitive Electrophysiological Testing*

Tests and Findings	Disease	Supporting Clinical Information
Normal ERG Abnormal PERG	Optic neuropathy	Optic pallor Characteristic visual field changes
VECP testing to localize laterality demonstrates increased chiasmal crossing of temporal fibers	Albinism (all types)	Iris transillumination Blond fundus/skin/hair (varies with form)
Nonrecordable to poor photopic ERG Normal to near normal rod ERG Normal visual field (except possible central scotomas) loss	Cone degeneration or dysfunction	Occasional temporal optic atrophy Macular atrophy, often concentric
Negative-wave, bright-flash, dark-adapted ERG Nonrecordable rod ERG Normal to subnormal photopic ERG Missing to subnormal oscillatory potentials Normal visual field	Congenital stationary night blindness, X-linked and autosomal recessive	No fundus findings Myopia Congenital night blindness
Negative-wave, bright-flash, dark-adapted ERG Subnormal rod and cone ERG Macular changes are characteristic Visual field is usually full	X-linked retinoschisis	Macular and peripheral schisis May present with vitreous hemorrhage
Abnormal/nonrecordable rod ERG (changes in b-wave sensitivity, V/V_{max}) Abnormal to borderline cone ERG Visual field loss (may be relative scotoma early)	Rod-cone degeneration	Pigmentary retinopathy Visual field loss Night blindness Family history
Abnormal/nonrecordable cone ERG Abnormal rod ERG (greater in amplitude than cone) Visual field loss will determine if in RP or cone degeneration category Final rod threshold elevation <2.2 log units	Cone-rod dysfunction[18, 19]	Temporal optic atrophy, telangiectatic optic nerve and adjacent retina Frequently less pigment deposits Occasional bull's-eye maculae
Normal ERG Abnormal EOG Dominant family history	Possible Best's disease Fundus and family examination will clarify if pattern dystrophy	Egg yolk lesion macula or symmetrical disturbance Visual acuity better than expected

*Mendelian inheritance pattern, course of the disease process, and fundus pattern often have to be utilized in addition to electrophysiological testing to determine a final diagnosis.

responses to the PERG may be abnormal early in the course of the disease.

Additional information about RPE/receptor disease can be obtained by performing both EOGs and ERGs on the same patient. In some disorders both results will be abnormal. In advanced and late stages of RP, both the ERG and the slow oscillation of the EOG will be markedly abnormal. However, in very early RP, the fast oscillations of the EOG may be more abnormal than the slow oscillation.

A distinctive pattern of abnormality that stands alone in electrophysiology is the abnormal misrouting of temporal retinostriate projections as detected by the VECP in oculocutaneous and ocular forms of albinism.[1] Testing must include measurements of cortical potentials over both the right and left occipital regions in order to obtain the required information. For reasons that are at present unclear, temporal retinal fibers that should pass to the ipsilateral visual cortex decussate in the chiasm in albinos to project to the opposite visual cortex. Thus, patients with albinism show a deficiency of ipsilateral or uncrossed responses. This pattern of abnormal retinocortical visual representation is so characteristic of all types of albinism that it is included in the operational definition as to which disorders should be called albinism and which represent only a variable hypopigmentation state from other causes.

The best stimulus to elicit VECR, for detecting asymmetry of retinocortical projections appears to be the onset/disappearance of large patterns against a homogeneous gray field of equal mean luminance rather than flashes or pattern reversal.[7] The best way of demonstrating the abnormal cortical projections is an examination for asymmetry of the left-minus-right occipital VECP signals as stimulated through the right and left eyes. This avoids problems with interpretation of hemispheric differences related to ocular dominance or individual differences in cortical topography.

Alternately, monocular VECP testing with bilateral occipital recording may demonstrate disproportionate uncrossed nerve fibers characteristic of all types of albinism.

CORRELATION OF TEST RESULTS WITH CLINICAL FINDINGS

There is often a misconception that individual electrophysiological tests are always diagnostic and can be interpreted in isolation without other tests and often without regard to the ocular findings. Where there are some characteristic electrophysiological tests for specific disease states, the vast majority of patients need a combination of tests selected to properly arrive at a diagnosis. A few patients will present who have extensive testing but in whom a diagnosis is not found. In order to better define these patients serial testing needs to be performed, often over years, in a search for diagnostic changes or for progression or stability in the disease; this allows for sensible counseling as well as placing the patient in a diagnostic category. Most patients accept the necessity to do serial testing if the rationale for evaluating the possibility of progression is explained.

Table 61–4 gives examples of ERG patterns that when correlated with the clinical findings are diagnostic of specific diseases or disorders.

It is also important to correlate the clinical findings with the electrophysiological and psychophysical data; otherwise a patient's condition may be misinterpreted. More frequently, the opposite occurs, where incorrect diagnoses are made because electrophysiological testing is not employed and clinical signs or symptoms suggesting a dystrophic or visual pathway disease are misinterpreted or neglected. Classic examples of difficult conditions to detect clinically but easy to diagnose electrophysiologically are RP sine pigmento, cone dystrophies with or without macular involvement, congenital stationary night blindness, Best's macular dystrophy, optic neuropathies without disc pallor, and hysterical amblyopia.

Kinetic visual fields correlating with the ERG and retinal findings often give a definitive answer as to the type of problem that a patient has; this is particularly relevant in cases of cone-rod dysfunction (with or without a bull's-eye macular lesion) in which the photopic b-wave is proportionately more affected than the rod signal but both are abnormal.

Patients with the cone-rod ERG pattern dysfunction may have a variety of problems, including a progressive disorder similar to RP, a cone degeneration with a subnormal rod response, occasionally juvenile retinoschisis (although the negative wave is present), advanced fundus flavimaculatus with severe macular loss, and cone-rod dystrophy.

A proportion of cone-rod dystrophies in which the cone ERG is disproportionately depressed follow a course similar to typical RP (rod-cone degeneration) and exhibit at some stage alterations of the peripheral visual field, often with a ring scotoma. Others progress clinically with a pattern of change like cone degeneration or dysfunction and have (until very late in the condition) full peripheral visual

fields and no ring scotomas, but may have central scotomas. One of the more useful aspects of serial visual field testing of many patients with retinal dystrophies, particularly RP, is to reassure the patient that the condition is not progressing rapidly. Anxiety levels are usually high in these patients, and annual fields showing little change reinforces the concept that the patient has a chronic disease and sudden blindness is very unlikely. A patient who is demonstrating rapid change should be carefully evaluated for the possibility of cystoid macular edema, hyperthyroidism, uveitis, serious systemic disease, and drug (or rarely light) toxicity. Occasional patients with RP will demonstrate reversible visual loss during episodes of viral or bacterial systemic or respirator infections. A few patients will complain of visual loss during pregnancy, but in most patients this appears to be temporary.[13]

Dark adaptation testing traditionally has been performed by measuring the threshold of vision after a preadapting bleaching with bright light. This must be done to standardize the test. Most laboratories use the Goldmann-Weekers dark adaptometer and a nonstandardized technique to determine the white light threshold of perception to a small light presented 10 degrees eccentrically. In normals, the threshold-time relationship is biphasic. If the preadaptation bleaches more than 50% of the rhodopsin, the scotopic system will take 10 minutes or more to become more sensitive than the photopic system. Therefore, after a rapid initial dark adaptation of 1 to 3 minutes, "cone" thresholds determine sensitivity until the cone-rod break, after which rods develop around 3.5-log-unit greater sensitivity. More sophisticated dark adaptometers are automated and test various points in the peripheral retina; they employ red and blue-green test objects so that the relative sensitivities of the scotopic and photopic systems can be determined at various times during dark adaptation.[9, 14]

In a number of patients with various types of retinal dystrophy, patients will relate a history of night blindness such that there are expectations when psychophysical testing is done to measure rod sensitivity that it should be very abnormal. Frequently, there is little correlation between final rod thresholds after 40 minutes of dark adaptation and the subjective complaint. Many patients with RP feel that their night vision is "fine" since it has not changed from childhood. It is useful to take measurements from at least two or three different retinal areas and take care to avoid known scotomas found on visual field testing. Patients with type I RP will typically have at least 3.5 log units of elevation of the final rod threshold, while patients with type II RP often have 2.5 log units or less. Type II can have more severe loss in more advanced stages where the visual field is less than 10 degrees.

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