Principles and Practice of Clinical Electrophysiology of Vision

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PART IX

Technical Issues

48

Technical Issues in Electroretinography

Michael A. Sandberg

The purpose of this section is to present techniques and methods of interpretation current in our clinic. Further consideration is given to the issues of interpreting variations within a single session and between visits in a given patient, as well as differences that may exist due to age, sex, refraction, ocular pigmentation, pupil diameter, and media changes.

LIGHT STIMULATION

Ganzfeld Stimulator

The standard light source for routine clinical ERG is a Ganzfeld or full-field stimulator that provides a (nearly) homogenous distribution of light over the central 120 degrees of the retina[22]; for retinal eccentricities >60 degrees, retinal illuminance falls as a consequence of decreasing apparent pupillary area, although this is compensated in part by the curvature of the retina and by reduced light absorption in the ocular media with eccentricity.[25] Full-field stimulation allows faster cone and slower rod components of the ERG to be separated in time and quantified not only with respect to amplitude but also with respect to implicit time.[3, 4] It also maximizes the reproducibility of consecutive responses in patients with variable fixation because the retinal light distribution is little altered by small changes in eye position. Full-field stimulation is provided by a large-diameter (e.g., 16 in.) dome, which may be fashioned from an unfinished plastic world globe (e.g., Fig 48–1).[34]

The dome is mounted on a table with adjustable height or have an adjustable chin rest so that the patient's head can be positioned vertically without discomfort. A red light-emitting diode is mounted in the rear of the dome opposite the head opening (Fig 48–1,F) to facilitate fixation and eye stability.

Different types of ERG contact lens electrodes have been proposed for full-field recording, some of which do not require an external Ganzfeld stimulator and circumvent obstruction of light by the brow and nose (i.e., lenses with corneal diffusers, wide-angle lenses, or built-in light sources). A theoretical study of retinal light distributions for small and large pupil diameters concluded that only lenses that were ≥12 mm in corneal diameter provided a light distribution that was nearly homogenous and minimally altered by pupil size.[22] Both clear and diffusing ERG contact lenses were reported to provide satisfactory light distributions,[22] although only with a clear lens would the patient be able to provide his own fixation and help maintain the pupil centered with respect to light incident on the corneal contact lens. Many ERG testers prefer that a lid speculum surround the ERG contact lens electrode to prevent the upper and lower eyelids from partially covering the cornea and thereby obstructing the passage of light into the eye; such reduction in retinal illuminance could reduce ERG amplitudes and increase implicit times. The lid speculum may also allow the tester to observe whether the patient's eyes are rolled up.

Effort should always be made to achieve as large a pupillary dilation as possible with a mydriatic agent in order to standardize the response and to maximize amplitudes for a given stimulus intensity. Response amplitude increases with increasing pupil diameter.[12, 21] However, the effect of change in pupil

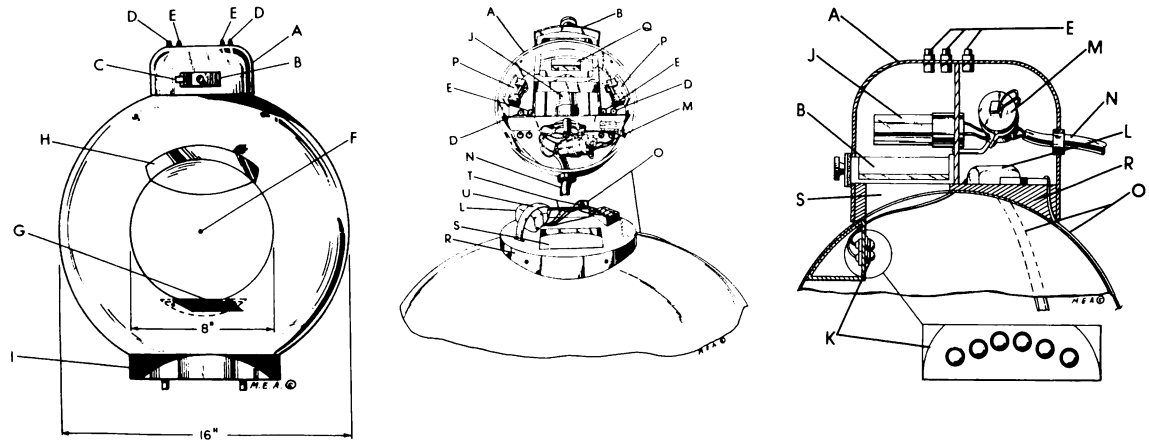

FIG 48–1.
Full-field (Ganzfeld) dome: *front, top,* and *side views. A* = flash housing; *B* = filter drawer; *C* = filter door latch (secures the drawer when the dome is mounted in the vertical position); *D* = lens input terminals, left and right channel (two channels allow recording from both eyes simultaneously); *E* = output to amplifier(s), left and right channel; *F* = central fixation light; *G* = chin rest; *H* = baffle; *I* = base; *J* = flash tube; *K* = background lights; *L* = socket to background light control; *M* = trigger coil; *N* = input from photostimulator chassis; *O* = wiring for fixation lights; *P* = fuses and fuse holders; *Q* = drawer retainer; *R* = mounting for flash housing; *S* = aperture for light entry; *T* = terminal strips for background lights and fixation light wiring; *U* = wiring to background lights. (From Rabin AR, Berson EL: *Arch Ophthalmol* 1974; 92:59–63. Used by permission.)

diameter may not simply be to alter retinal illuminance. A study of the full-field ERG in rabbits[12] found that constriction of the pupil from 10- to 5-mm diameter had a large effect on amplitude but little effect on implicit time for the rod ERG. In other words, the effect on the ERG of reduced pupillary aperture could not be compensated for by increased light intensity. As a rule, ERG testing should only be done in patients with dilated pupillary diameters ≥6 mm.

Flash Illumination

Routine full-field ERGs are commonly elicited by 10-μs-duration flashes from a xenon flash lamp (e.g., Grass Instruments) positioned above an external dome (Fig 48–1,J). It may be convenient to mount the flash lamp within its original housing to facilitate replacement of aged lamps or for exchange with a photoflash unit that increases the maximum integrated retinal illuminance by ~3 log units in order to elicit oscillatory potentials or responses from eyes with opaque media. The light flashes are attenuated by gelatin neutral-density and/or chromatic filters (e.g., Eastman Kodak Co) positioned beneath the flash lamp (Fig 48–1,B) and then diffused by glass, plastic, or paper before entering the aperture for light entry (Fig 48–1,S). A plastic strip should be positioned between the place where the light enters the dome and the subject's head (Fig 48–1,H) to prevent direct stimulation of the eye. The interior surface of the Ganzfeld dome should be coated with a matte white (smoked magnesium oxide or barium sulfate) paint (e.g., Eastman Kodak Co) to provide a nonspecular, spectrally flat light distribution.

Background Illumination

Background illumination is used to desensitize rods and isolate the cone ERG.[4] Typically, this is provided by an array of tungsten filament lamps (Fig 48–1,K) connected in parallel to a dc power supply and mounted, facing the diffuser, on the same plastic strip used to mask direct view of the stimulus from the eye. Alternatively, background lighting can be obtained from a bifurcated fiber optic illuminated by a high-intensity tungsten-halogen lamp, a fiber bundle entering the dome to each side of the front opening; in this case, the fiber optics need to be equipped with diffusers. The fiber optics make it convenient to attenuate the background illumination by neutral or colored filters. Tungsten illumination makes it possible to raise the background illumination gradually with the aid of a rheostat to prolong the life of the lamps. The rheostat is also important for testing patients with hereditary retinal disease who may be photophobic and find sudden onset of the background aversive. If background illumination is altered in the protocol, calibration of the luminance should be done prior to testing. A small variance in background can greatly affect the ERG.[3, 27]

Focal Stimulation

Several light stimulators have been developed for eliciting focal cone ERGs from the retina, primarily for the purpose of detecting foveal malfunction that is not evident in full-field recordings.[2] Most focal stimulators present flashes in the presence of a photopic background or surround that desensitizes both rods and cones adjacent to the stimulus field.[7, 19, 38] The flashes should be white,[38] rather than red (as has been generally used), to avoid eliciting subnormal amplitudes from patients with congenital protanopia. The most important feature for eliciting focal cone ERGs is a means to limit light stimulation to a given region of the retina (e.g., the fovea). If the stimulus were inadvertantly positioned on an area eccentric to the fovea, for example, in patients with strabismic amblyopia with eccentric fixation, a low-amplitude and/or slow response that might be normal for that eccentric region would be interpreted as subnormal and/or delayed if thought erroneously to be generated by the fovea. In patients with suspected macular disease who may have reduced visual acuity and eccentric and/or variable fixation, the conventional method of having a patient fixate on the center of the stimulus should not be relied upon. An alternative approach has been to visualize the stimulus ophthalmoscopically on the fundus and position the stimulus on the area of interest throughout testing.[19, 38] In this approach, the background or surround not only serves to desensitize photoreceptors outside the stimulus patch but also allows identification of retinal landmarks to facilitate localization of the stimulus on the area of interest. In a prototype instrument (Fig 48–2), the stimulus and background beams originate from a single lamp (Fig 48–2, no. 1) and later merge at a beam splitter (Fig 48–2, no. 24) to enter the patient's dilated pupil coaxially in maxwellian view.[36]

These features allow the relative luminous efficiencies of the stimulus and background beams to remain substantially constant, even with variation in the angle of pupillary entry, to minimize variability in the cone ERG. The device is hand holdable, so the examiner can easily compensate for slow changes in the patient's head or eye position and thereby track the retinal area being tested. A stimulator for eliciting focal rod ERGs has not yet been reported.

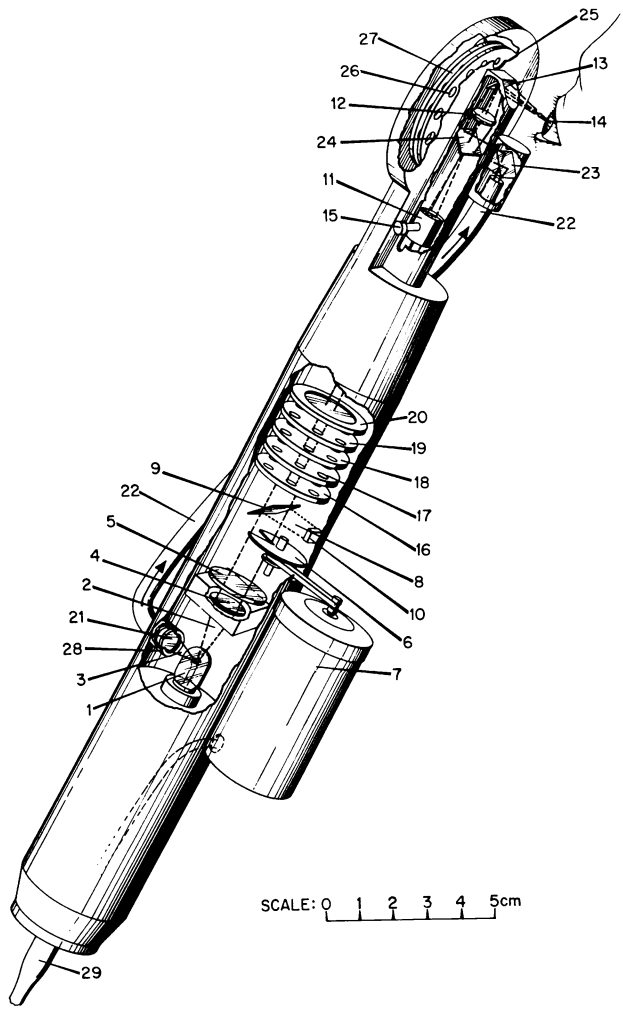

FIG 48–2.
Hand-held, dual-beam, maxwellian-view stimulator-ophthalmoscope. Symbols designate the following: *1* = incandescent lamp for both stimulus and background channels; *2* = stimulus channel; *3* = background channel; *4* = collimating lens; *5* = heat-absorbing glass; *6* = rotating sector disk; *7* = motor; *8* = reflected stimulus beam; *9* = beam splitter; *10* = photocell; *11* = reducing lens; *12* = focusing lens; *13* = first-surface mirror; *14* = patient's pupil; *15* = knob; *16–20* = rotatable discs to adjust stimulus intensity, wavelength, diameter, and/or polarization; *21* = heat-absorbing glass; *22* = fiber optic; *23* = reflecting prism; *24* = beam splitter; *25* = viewing aperture; *26* and *27* = rotating lens assemblies to bring the fundus into focus; *28* = filter housing; *29* = power cord. (From Sandberg MA, Ariel M: *Arch Ophthalmol* 1977; 95:1881–1882. Used by permission.)

Light Calibration

Calibration of stimulus and background luminances may be done with a portable, battery-operated electronic photometer (e.g., United Detector Technology). The instrument can measure directly in illuminance (i.e., light falling on a surface) in units of foot-candles, luminance (i.e., as seen by the subject) in units of foot-lamberts (ft-L), or in integrated luminance in units of foot-lambert-seconds by

taking into account flash duration. For a Ganzfeld dome with xenon flashes of light, the most convenient way to monitor luminance calibration periodically is to place the p-i-n–photodiode, photopic filter, and lens assembly on the chin rest, set the repetition rate to 30 Hz, and measure the steady-state luminance; single-flash luminance may then be calculated by taking into account the ratio of light divided by light + darkness. Calibration may also be done by summing consecutive flashes as integrated luminance, which is then divided by the number of flashes to obtain a mean value, or as luminance by also taking into account the flash duration; this is the only applicable method for calibrating each of the five intensity settings of the photostimulator (i.e., 1, 2, 4, 8, and 16), since intensity falls for the higher settings when the stimulus is flickering, or for calibrating a photoflash unit with a recycling time of 15 to 30 seconds. Calibration of background luminance can be done directly as a steady-state value.

Preparation of Subject for Electroretinographic Testing

ERG testing is done first under conditions of complete dark adaptation and afterward under conditions of light adaptation because this sequence easily keeps lens wearing time below a maximum of 45 minutes to minimize the risk of a corneal abrasion and facilitates obtaining reproducible, maximal responses because the eye light-adapts faster than it dark-adapts. Subjects should have their pupils maximally dilated (e.g., 2 or more drops of 2.5% phenylephrine hydrochloride and 1% cyclopentolate hydrochloride) and then should be dark-adapted for at least 45 minutes. At least two sets of drops of dilating agent are recommended because a patient may blink out one or more drops and pupillary dilation could be inadequate.

Since we use eye patches to occlude ambient light to dark-adapt patients, the patient is then led from the waiting area into a darkened testing area by the technician who wears a battery-powered headlamp attenuated by a red gelatin filter (e.g., Eastman Kodak Co); the red filter provides dim photopic illumination for the technician and, with indirect illumination of the eye, will not light-adapt the patient. A silver cup electrode containing electrode cream as ground is placed on the forehead and secured with tape, with the wire led off to the side of the head and its plug inserted into an electrode junction box. The patches are then removed from one eye and 2 to 3 drops of proparacaine hydrochloride instilled in the lower fornix. The pupil should then be inspected for adequate dilation and redilated, if necessary. With the patient looking down, the technician then raises the upper lid with his forefinger or thumb and inserts the upper speculum of a bipolar Burian-Allen contact lens electrode (e.g., Hansen Ophthalmic Development Laboratory) containing 1 drop of methylcellulose beneath the upper lid. With the patient looking straight, the technician then pulls down the lower lid in like manner, inserts the lower speculum, and promptly secures the lens cable to the cheek with slack by tape to avoid tugging on the lens by the patient accidentally brushing against the remainder of the lens cable. The lens leads should then be inserted into the junction box. Bipolar Burian-Allen lenses can be obtained with conventional stalks for easy handling for use in full-field stimulation or with specialized short stalks for use in focal stimulation with the stimulator-ophthalmoscope in order for the examiner to focus its beams in the plane of the pupil (i.e., in maxwellian view). The contact lens should be centered over the pupil by adjusting the length or position of the lens cable fixed by tape to the cheek or, if necessary, by affixing tape to the upper speculum (or lens stalk). If tape is used across the speculum, it should be positioned so as not to interfere with the passage of light to the pupil. The technician should confirm, both before and after testing, that the upper and lower speculums are entirely beneath the upper and lower lids, respectively; otherwise, illumination of areas of the retina could be obstructed by the lid and subnormal amplitudes and delayed implicit times obtained, even without significant electrical noise.

For full-field stimulation, the subject's head is placed on the chin rest (Fig 48–1,G) and projecting within the dome so that the eye to be tested cannot be visualized by the examiner from the side (i.e., the dome fills 180 degrees of the visual field). The subject is then asked to look at the fixation spot and determine whether he sees one or two spots. If two spots are seen, the contact lens is probably centered below the line of vision and should be raised to optimize retinal illuminance and electrical contact. For focal stimulation with a hand-held stimulator-ophthalmoscope, the patient should be reclined and the head turned toward the examiner, who is seated. The examiner then directs the maxwellian-view beams through the patient's dilated pupil and focuses the lens assembly (Fig 48–2, nos. 26 and 27)

for clear visualization of the macula. A critical aspect of focal ERG testing is confirmation by the examiner that the stimulus can be clearly visualized and is properly positioned on the fundus. If a double image of the stimulus is seen on the retina, then the contact lens has an air bubble that needs to be eliminated. The light beams should be directed as nearly as possible through the center of the contact lens and pupil to avoid a reduction in retinal illuminance by the electrode ring or iris. The appearance of a specular reflex with scattered light off the retina can be avoided by a small shift in instrument position. If clear visualization or localization of the stimulus on the fundus by the examiner cannot be obtained as, for example, in patients with corneal opacities, poor dilation, or significant cataracts, or in patients with large deviations in eye position, then focal stimulation of a given area cannot be confirmed and should not be attempted. Following testing, the contact lens should be cleaned by ultrasound with a cleaning solution, rinsed in water, and air-dried.

Dark and Light Adaptation

Full-field rod b-wave amplitude increases during dark adaptation. Therefore, patients should be initially dark-adapted to obtain maximal, stable amplitudes. The time in the dark required to achieve a maximal ERG depends upon the prior light exposure and may require 45 minutes or longer, even following exposure to room illumination, as shown for a normal subject following a 10–ft-L bleach (Fig 48–3).

Even 1 hour of dark adaptation may be insufficient for a 500–ft-L bleach (Fig 48–3), which is only ~10% of the light used in fundus examination. Conversely, full-field cone ERG amplitude increases during light adaptation in normal subjects[28] and some patients with retinal disease.[29] Maximum amplitudes may require 5 minutes or longer of light adaptation, and amplitude stability should then be confirmed in consecutive recordings spanning 2 to 3 minutes (Fig 48–4).

Focal cone ERGs can be performed in darkness for optimum visualization of the fundus or in dim ambient room illumination. It has not yet been determined whether focal cone ERG amplitude varies systematically during light adaptation, although consecutive response averages recorded over a period of a few minutes have been found to be reproducible in all patients so far reported.

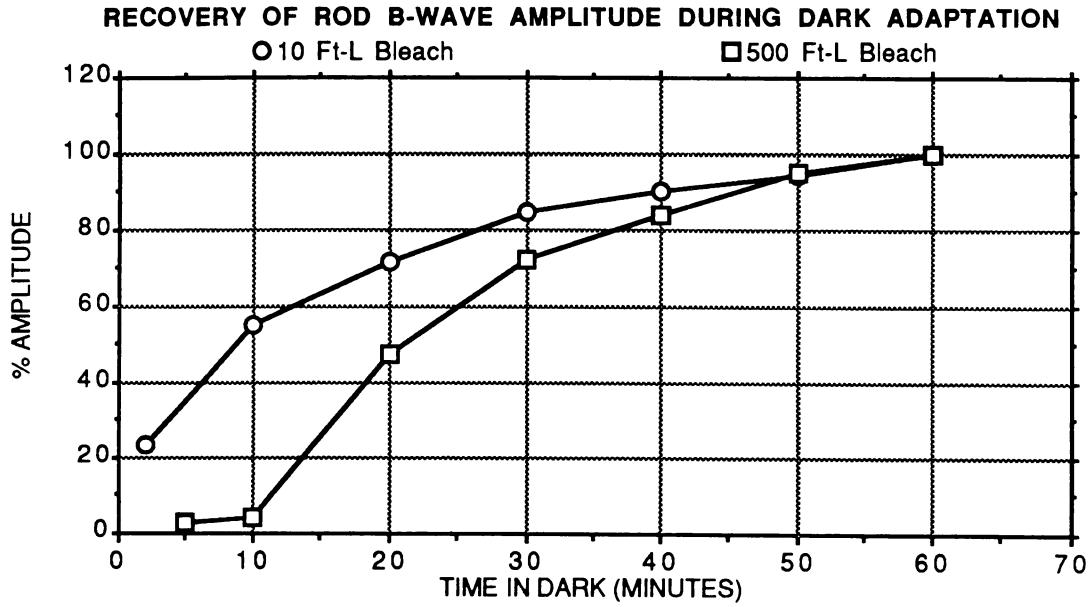

FIG 48–3.

Computer-averaged (n = 16) full-field rod ERG percent amplitudes to an 0.5-Hz, 10-μs blue flash (λ_{max} = 440 nm) of 16 ft-L recorded over time in the dark from a normal observer after a 10-minute, full-field, white light bleach of 10 or 500 ft-L presented to the dilated pupil. Amplitudes for 60 minutes' dark adaptation were arbitrarily designated 100%. All the data appear to reflect rod function, except for the low-amplitude 5-minute value after the stronger bleach, which probably represents residual cone function. Each set of data represents a single run.

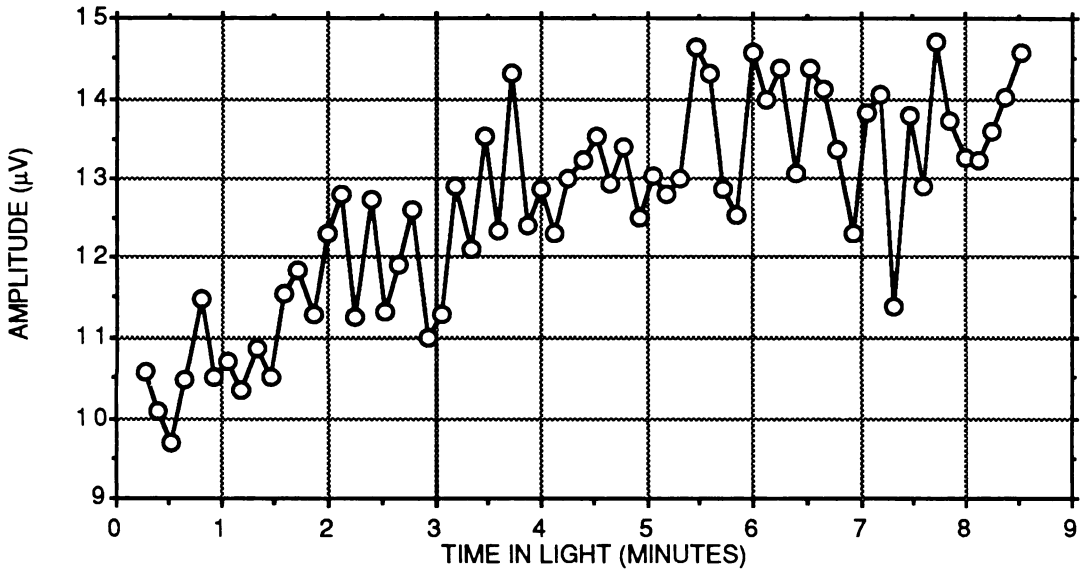

FIG 48–4.
Computer-averaged (n = 32) cone ERG amplitudes recorded at different times during presentation of 30-Hz, white, full-field, 10-μs flashes of 6,300–ft-L luminance through the dilated pupil of a previously dark-adapted patient with dominant retinitis pigmentosa *(RP)* with reduced penetrance.

NORMAL VARIATION

The variation in full-field ERG amplitude for a given stimulus condition across normal subjects is as much as 2.5:1.[6, 27] Several factors have been identified that appear to contribute to this normal variation, as follows.

Effect of Age

For example, it is well known that b-wave amplitude decreases with age among adults.[27, 33, 41, 43] A study of 24 normal subjects between the ages of 9 and 67 years with low refractive errors found that the linear regressions of full-field b-wave amplitude on age had slopes between −2.1 and −2.6 μV/yr for a variety of stimulus conditions (e.g., Fig 48–5)[41]; these values were comparable to some reported in another study.[27]

Approximately 30% of the variance in b-wave amplitude for all the subjects combined could be explained on the basis of age.[41] However, no significant age dependencies were seen for a-wave amplitudes (except for a small slope for the light-adapted cone a-wave) or for a-wave or b-wave implicit times, although another study found the a-wave amplitude to fall with increasing age[27] and two other studies found b-wave implicit times to increase with increasing age.[27, 43] The cone b-wave implicit time to a flickering white light is known to be longer in patients above 60 years of age than in younger patients. Some of the reduction in amplitudes and increase in implicit times among the elderly for submaximal responses are probably due to reduced light transmissivity by the lens[35] and, possibly, to a decrease in maximum pupil diameter from mydriasis with increasing age.

In contrast to the inverse relation between ERG amplitude and age among adults, ERG amplitude has been reported to increase with age among young children; this was based on 126 children tested under general anesthesia between the ages of 3 months and 8 years (Fig 48–6).[14]

Most of the change was observed before 1 year of age; therefore, it would be best to test children after 1 year of age, when amplitudes would be expected to have stabilized, in order to assess normality of the ERG. Cone and rod b-wave implicit times were found to decrease with increasing age,[14] in contrast to what has been reported for adults.[27, 43] An age dependency has also been reported for amplitude in the focal cone ERG. Foveal cone ERG amplitude has been reported to be inversely correlated with age for 100 normal eyes of subjects between the ages of 5 and 75 years, with a slope of −.002 μV/yr.[9] Cone b-wave implicit time increased with increasing age.[9]

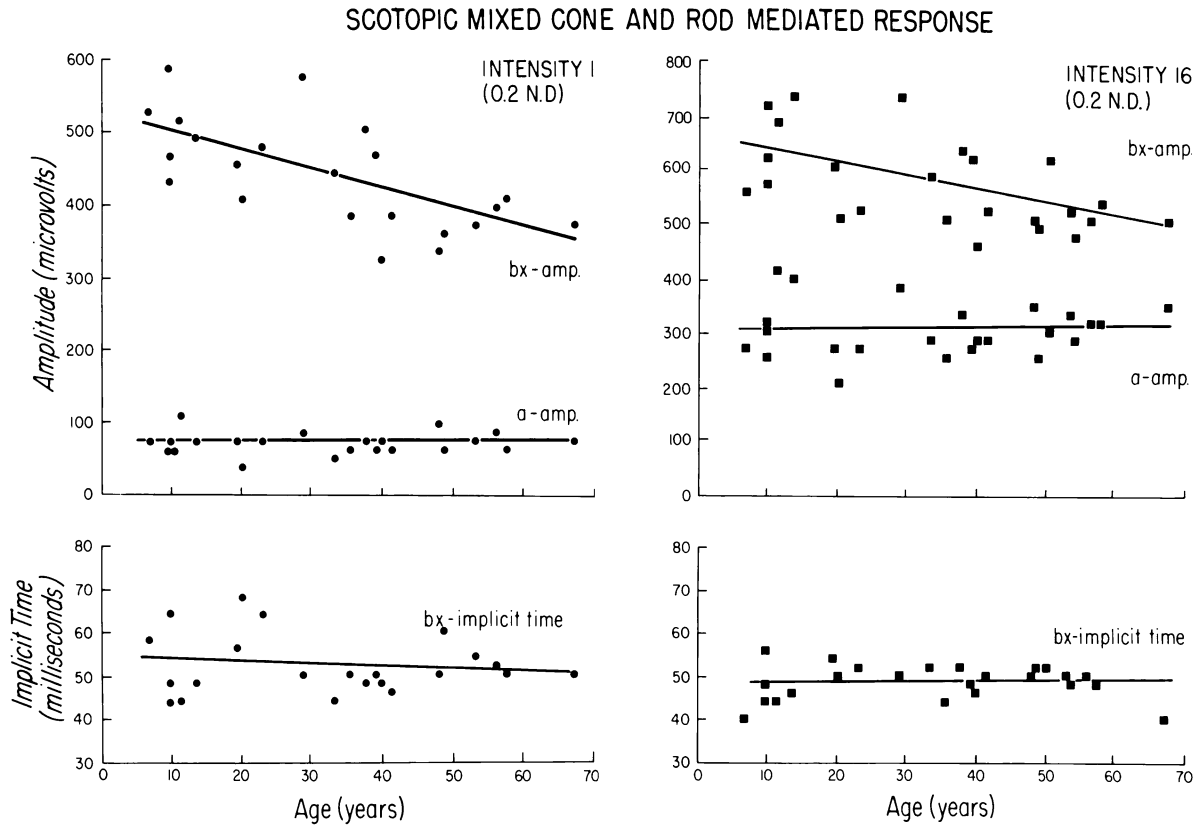

FIG 48–5.
Scattergrams for scotopic responses mediated by mixed cone and rod activity. Only the bx amplitude showed significant linear regression slopes. (Adapted from Weleber RG: *Invest Ophthalmol Vis Sci* 1981; 20:392–399. Used by permission.)

Effect of Sex and Refraction

The ERG b-wave has been consistently found to be higher in women than in men of comparable age.[20, 27, 31, 33, 45] On average, the difference is 30 to 40 μV for maximal responses. The amplitude of the b-wave has been also reported to vary with the refraction of the eye, being smaller in myopic and larger in hyperopic eyes (Fig 48–7).[31–33]

The relevant refractive component appears to be axial length of the eye since b-wave amplitude in normal subjects varies inversely with axial length.[31, 32] In fact, the dependencies of b-wave amplitude on sex and refraction may both reflect a proportionality between b-wave amplitude and 1/tissue resistance between recording electrodes, as found between corneal and forehead electrodes in 19 young adult normal observers with small refractive error (Fig 48–8).[26]

Whether a similar amplitude dependence on tissue resistance can be shown for a bipolar ERG contact lens in which the geometric relation between positive (corneal) and reference (speculum) electrodes is fixed remains to be determined. If electrode resistance varies directly with axial length in humans, then the smaller axial lengths in women[31] and in hyperopes could account for their larger b-wave amplitudes.[31] A study of electrode resistance for a bipolar Burian-Allen contact lens vs. axial length should help to resolve this issue.

Effect of Ocular Pigmentation

Larger b-wave responses have been reported for albinos as compared with whites,[21] for whites as compared with blacks,[15] and for albinos as compared with whites and carriers of ocular albinism.[23] However, these studies did not use full-field illumination of the retina, so the higher amplitudes observed in eyes with lighter pigmentation were probably due, at least in part, to increased illumination of indirectly stimulated areas. On the other hand, one of the studies found increased maximal b-waves for albinos,[23] thus suggesting that another contributing factor may also be involved.

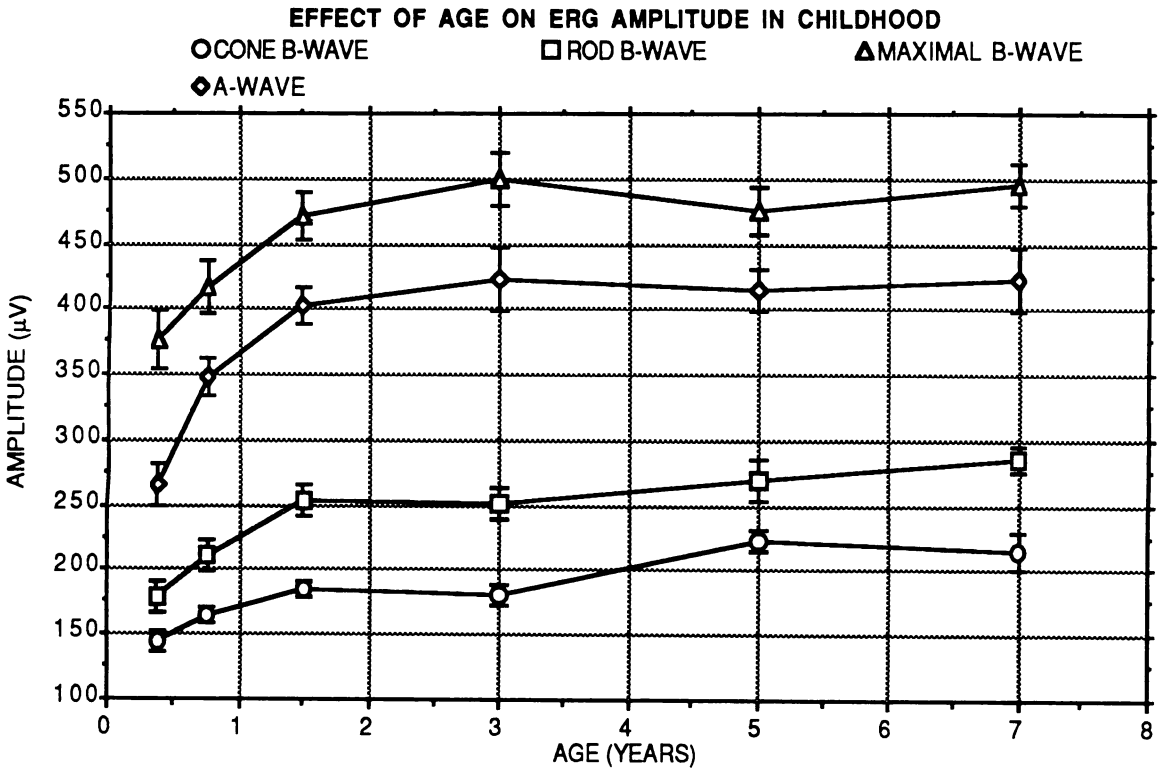

FIG 48–6.
Mean ERG amplitudes and standard errors vs. age in 126 children tested under halothane anesthesia. Cone b-waves were elicited under conditions of light adaptation, rod b-waves were elicited with a blue stimulus of 1 J, maximal b-waves were elicited under scotopic conditions, and a-waves were elicited with a stimulus of 10 J. (Data from De Rouck A: *Bull Soc Belge Ophtalmol* 1982; 202:97–113.) The author also evaluated children of some of these ages and of older ages without general anesthesia, but the results of these measurements are not presented here.

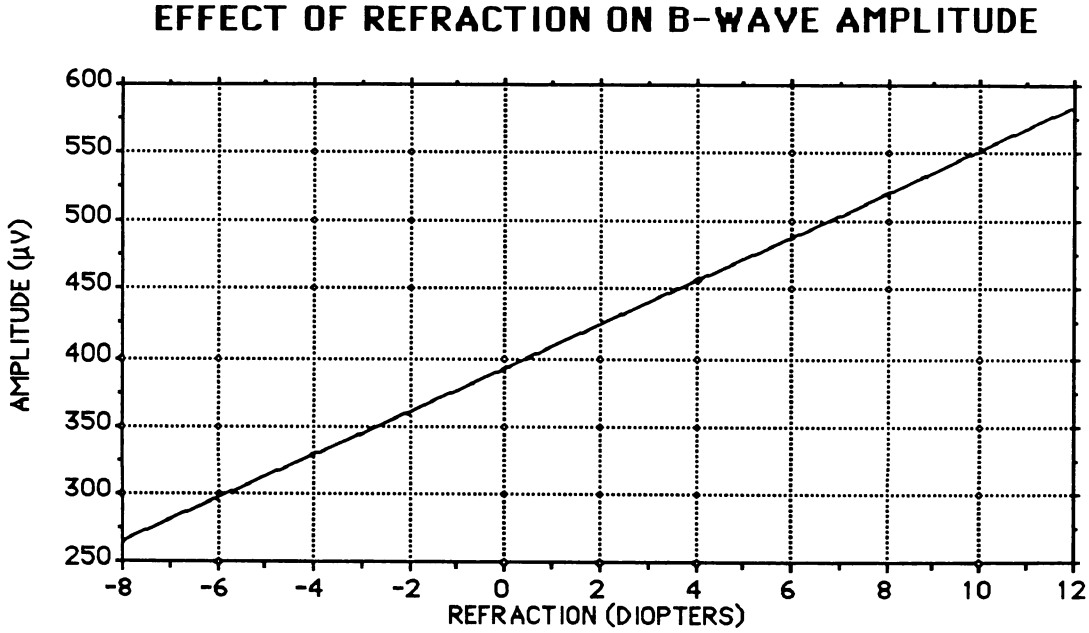

FIG 48–7.
Linear regression of b-wave amplitude on refraction based on the average of regression equations for men and women from measurements on 86 normal subjects. Maximal b-waves were elicited from the semi–dark-adapted eye with short, intermittent white light flashes ranging from 20 to 800 lux derived from a lamp at different distances from the subject and presented through the dilated pupil. (Data from Pallin E: *Acta Ophthalmol.* 1969; 101(suppl):1–57.)

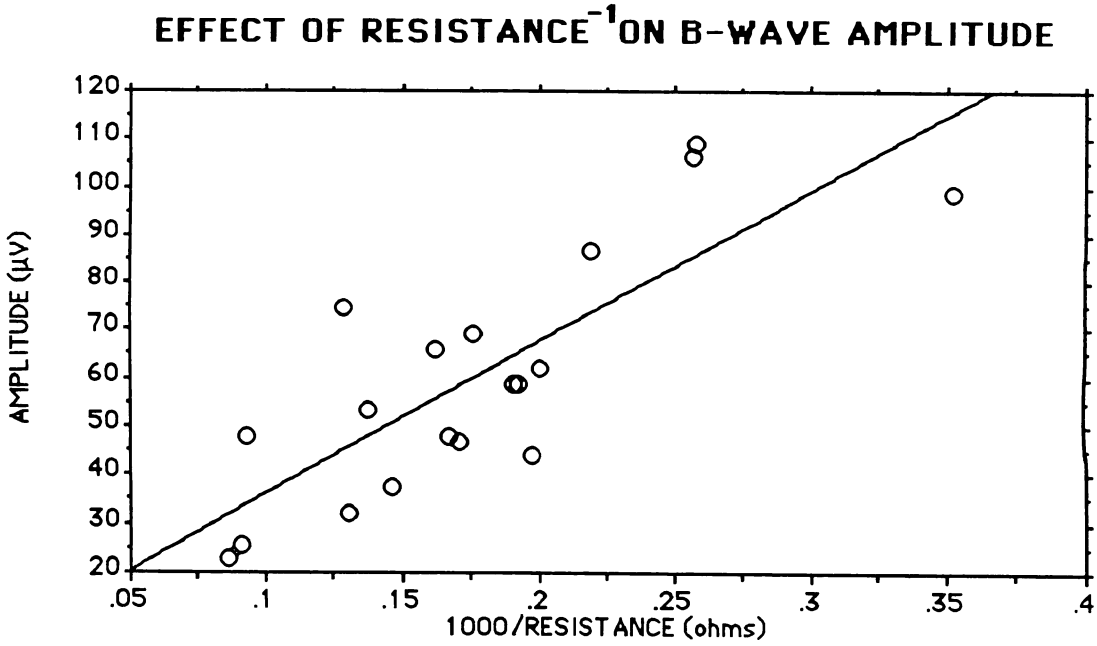

FIG 48–8.
Linear regression of b-wave amplitude on inverse resistance for 19 normal subjects between the ages 25 and 40 years and with refractive errors between −1.50 and +1.25 D. ERGs were elicited with Grass PS22 photostimulator flashes at intensity 2 illuminating a corneal diffuser from a distance of 50 cm. Responses were monitored with a contact lens electrode referenced to a silver electrode on the forehead midline and a ground electrode on the earlobe. Resistance was measured by passing a 250-Hz, 0.1-μA current between the corneal and forehead electrodes. (Data from Lemagne J-M, Gagné S, Cortin P: *Can J Ophthalmol* 1982; 17:67–69.)

CHANGES IN OCULAR MEDIA

Effect of Corneal Opacity, Miosis, Cataract, and Vitreous Hemorrhage

Patients who are taking miotics for glaucoma or who have posterior synechiae or other pupillary obstruction will likely have a pupillary diameter <6.0 mm after mydriasis and ERGs that are smaller in amplitude and often longer in implicit time than expected due to reduced stimulus retinal illuminance. Patients with corneal opacities, large cataracts, or vitreous hemorrhages that obscure visualization of the fundus will also likely have their ERGs reduced in amplitude and slowed in implicit time because the stimulus retinal illuminance will be reduced when compared with that in eyes with clear media; responses may even be nondetectable with conventional full-field recording with a photostimulator. Alternatively, an electronic photoflash, with more than 10,000 times the energy of the conventional full-field flash when positioned a few inches from the eye, has been used to stimulate eyes with opaque media in order to generate sufficient retinal illumination to elicit a measurable ERG.[18] In order to separate the optical density effect of the media obstruction from any change that may be due to a retinal abnormality, responses to a series of stimulus intensities should be compared with those obtained with conventional full-field flashes in normal eyes. If a-wave and b-wave amplitudes and implicit times to the brighter flashes in eyes with opaque media can be matched to those obtained to the dimmer flashes in normal eyes with clear media, then large areas of the retina are functional. Note that a great degree of maculopathy can occur and not be detected by this technique.[24] This comparison would be facilitated if the photoflash were positioned over a Ganzfeld dome so that all areas of the retina were directly and comparably illuminated by the flash. A full-field bright-flash stimulator, although dimmer than a directly viewed bright-flash stimulator, still generates ~1,000 times the energy of the conventional full-field system, which should be sufficient to evaluate most patients.

Effect of Vitreous Substitute

Replacement of the vitreous with silicone oil during vitrectomy in patients with retinal detachment has been associated on follow-up with a marked reduction in ERG amplitude even in eyes in which the

retina had reattached[24] and where subjective measures of vision had improved.[13] Removal of silicone after reattachment of the retina in some patients was associated with an immediate recovery of the ERG, thus indicating that the loss of amplitude with silicone was due, at least in part, to a reduction in electrical conductivity of the ocular media by the insulating effects of silicone.[17, 30] Experimental[30] and theoretical[16] studies have reported that the percent reduction in ERG amplitude is nonlinearly proportional to the relative amount of vitreous replaced by silicone. Replacement of the vitreous with other nonconductive materials (e.g., gas) would be expected to result in similar attenuation of the ERG. Therefore, ERG assessment of retinal function following vitreous replacement should be made with caution.

INTERSESSION VARIABILITY

The ERG has been repeatedly recorded in normal persons and patients, to determine test-retest variability. This varies with the stimulus for small responses, variability of ± 50% can be seen. For large responses, variability of ± 25% can be seen.

Normal subjects not entrained to a cyclic pattern of illumination have shown no significant variation in rod ERG amplitude over daylight hours.[10] However, in normal subjects entrained to cyclic illumination for at least 3 days, rod ERG amplitudes follow a regular pattern, being on average 10% to 15% smaller 1.5 hours after light onset than at other times of day (Fig 48–9).[10, 11, 37]

Patients with dominant[37] or other types of retinitis pigmentosa[8] who are similarly entrained to cyclic

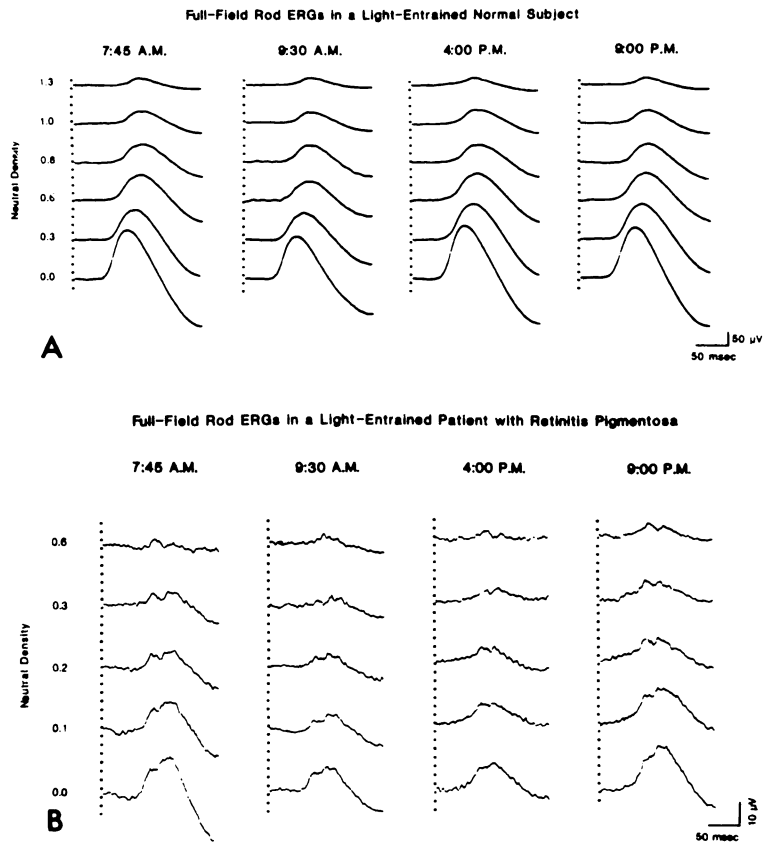

FIG 48–9.
Computer-averaged rod ERG responses (n = 16) obtained from the light entrained eye of a normal observer **(A)** and a 26-year-old patient with dominant retinitis pigmentosa **(B)** on a single day of testing. The subjects were entrained to an 8 AM light: 10 PM dark schedule. The stimulus flash was attenuated by neutral density filters as shown on *left*. The unattenuated integrated retinal illuminances were 0.0 log scot td-sec **(A)** and 0.4 log scot td-sec **(B).** Stimulus onset is indicated with *vertical broken lines.* (From Sandberg MA, Baruzzi CM, Hanson AH III, et al: *Invest Ophthalmol Vis Sci* 1988; 29:494–498. Used by permission.)

light have been shown to have an abnormally large variation in rod b-wave amplitude over the course of the day in their entrained eyes that amounts to as much as a 40% to 50% reduction 1.5 hours and/or 8 hours after light onset (see Fig 48–9). Since diurnal variation has not been investigated in nonentrained patients, we do not yet know if it is necessary to control for time of day in recording.

The physiological basis for these ERG diurnal rhythms may be associated with rod outer segment disc shedding since recordings in rats have demonstrated an ERG variation similar to that seen in humans which is significantly correlated with the number of phagosomes appearing in the retinal pigment epithelium.[39] Furthermore, this ERG rhythm is found even in Royal College of Surgeons (RCS) rats with hereditary retinal degeneration and a defect in phagocytosis, thus indicating that it is probably outer segment disc shedding and not engulfment of shed packets of discs that is associated with the reduction in ERG amplitude.[40] Changes in the rod b-wave have also been related to outer segment disc shedding in the rabbit.[42] In both humans[8, 10] and rats[39] the ERG variation has been comparable in the a-wave and b-wave, further implicating the photoreceptors in its generation.

PROBLEMS WITH BURIAN-ALLEN ELECTRODES

Intermittent lens contact may occur in subjects whose corneal curvature closely matches that of the contact lens electrode, whose upper or lower lid does not completely retain the speculum, or whose lids squeeze on the speculum. These eyes probably do not maintain a continuous bridge of electrolyte coating between the cornea and lens electrode. Intermittent contact may also result from a bend in the spring that holds the lens against the cornea. The sign of intermittent contact is occasional high-frequency noise, often present in some tracings and not in others, that may reduce response amplitude. When present, the noise can usually be eliminated by reinserting the contact lens with additional coating agent and fixing the lens in position by tape or by changing lenses.

A complete loss of lens contact, associated with popping out of the entire speculum from beneath the lids, is likely to occur in children, in patients with exophthalmos, and in patients who cannot visualize the red fixation spot and whose eyes may therefore undergo occasional extreme vertical deviations; a loss of contact may also occur as a result of a

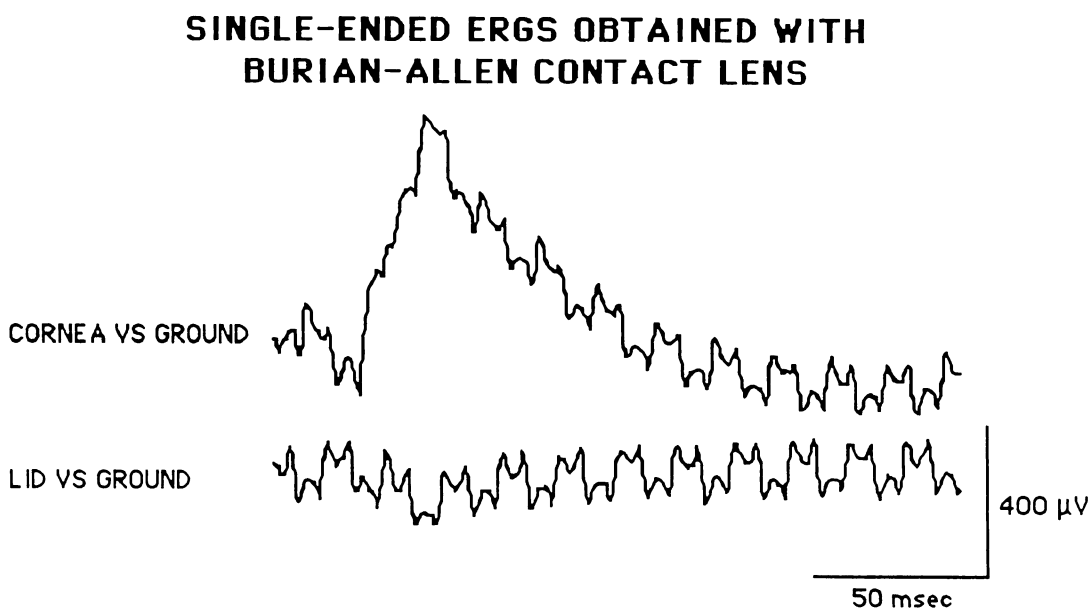

FIG 48–10.
Full-field responses from a normal subject wearing a bipolar Burian-Allen contact lens to 0.5-Hz, 10-μs white flashes of 6,300 ft-L presented through the dilated pupil to the dark-adapted eye for two monopolar recording configurations: cornea (i.e., positive electrode) vs. ground and speculum (i.e., negative electrode) vs. ground. The speculum potential appears as a low-amplitude response inverted by the amplifier. Line-frequency (i.e., 60-Hz) noise, superimposed on each monopolar response, appears nonsinusoidal due to amplifier saturation.

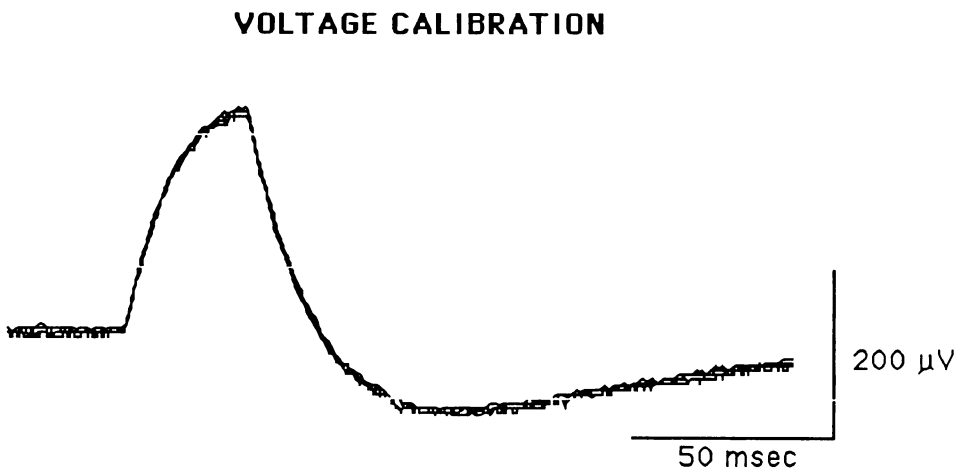

FIG 48–11.
Voltage calibration obtained with an electronic calibrator (LKC Instruments) triggered by light. Four consecutive waveforms have been superimposed to illustrate reproducibility.

break between a lens cable and its pin plug or due to a blown fuse. A loss of contact will result in line-frequency noise filling the oscilloscope or computer screen. With a broken lead, distorted line-frequency noise will be superimposed on a single-ended response (Fig 48–10).

Calibration Procedures

At the beginning of each day of testing and after sufficient time for equipment warm-up and stabilization, a time-varying calibrated voltage is put through the recording system to confirm normal function. It is desirable that this waveform resemble in amplitude and timing a normal ERG (e.g., Fig 48–11) so that amplifier, oscilloscope, and/or computer settings do not have to be changed for subsequently testing patients.

Nine times out of ten a problem will be one of electrical contact involving the ERG lens or one of its cables or plugs. The technician should always have accessible a shorting cable to connect the positive and negative inputs of the ERG lens junction box to determine whether the origin of the noise is between the patient and junction box or between the junction box and oscilloscope or computer; if the trace then becomes perfectly clean, the problem involves the ERG lens or one of its plugs. Some difficulties will stem from user error, such as an improperly adjusted trigger threshold or phosphor brightness, but only very rarely from equipment malfunction. User errors are best detected and corrected by having a written manual that defines the position of all knobs and switches. Equipment malfunction may be most conveniently circumvented by using modular components that can be quickly replaced by spare components.

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