Principles and Practice of Clinical Electrophysiology of Vision

Editors

JOHN R. HECKENLIVELY, M.D.
Professor of Ophthalmology
Jules Stein Eye Institute
Los Angeles, California

GEOFFREY B. ARDEN, M.D., PH.D.
Professor of Ophthalmology and Neurophysiology
Institute of Ophthalmology
Moorfields Eye Hospital
London, England

Associate Editors

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Chiba University School of Medicine
Chiba, Japan

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Department of Vision Sciences
Aston University
Birmingham, England

SVEN ERIK NILSSON, M.D., PH.D.
Professor of Ophthalmology
University of Linköping
Linköping, Sweden

RICHARD G. WELEBER, M.D.
Professor of Ophthalmology
University of Oregon Health Science Center
Portland, Oregon

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Log (bp/bs) in Retinitis Pigmentosa

Hiroyuki Iijima

Retinitis pigmentosa (RP) is a set of diseases mainly affecting photoreceptors and retinal pigment epithelium,[6] with a variable distribution of rod and cone involvement, which in patients with RP and allied diseases can be evaluated both psychophysically and electrophysiologically. Massof and Finkelstein[7] studied rod and cone function in RP patients with psychophysical sensitivity measured with two-color stimuli and proposed a new, now widely accepted RP classification based on varying involvement of the rods and cones. Electrophysiologically, rod and cone functions can be evaluated by measuring the scotopic (rod) and photopic (cone) electroretinographic (ERG) amplitudes.[3] Arden and associates[1] classified patients with autosomal dominant RP into two categories based on the presence or absence of the scotopic ERG. But it is not a consistent classification because their subgroup A, in which no scotopic ERG was evoked, could have had recordable scotopic ERGs in the early stages and their subgroup B, in which measurable scotopic ERGs were evoked, might show abolished responses several years later. Since both rods and cones are affected in most progressive photoreceptor diseases, rod vs. cone involvement can be more effectively evaluated by the ratio of rod and cone ERG amplitudes than by the amplitudes themselves.

We studied the scotopic (rod) b-wave amplitude (bs), the photopic b-wave amplitude (bp), and their quotient (bp/bs) in standardized clinical ERG recordings of normal subjects and patients with RP.[4] Since bp/bs appeared to follow a log normal distribution in normal subjects (Fig 65–1), log (bp/bs) was considered to be the most appropriate index for the evaluation of rod vs. cone involvement. Log (bp/bs) showed no relationships with either sex or age.[4]

The distribution pattern of log (bp/bs) in patients with autosomal dominant RP disclosed that the disease consists of at least two different categories, type 1 and type 2 (Fig 65–2). Type 1 patients had log (bp/bs) values larger than 0.5, while type 2 patients had log (bp/bs) values between −0.75 and 0.5. Type 1 patients had higher final dark adaptation threshold values and more delayed photopic b-wave implicit times. In this study the patients from the same family tree belonged to the same group. Our ERG classification of autosomal dominant RP broadly conformed to the psychophysical classifications proposed by Massof and Finkelstein[7] and by Lyness and associates.[5] These studies showed that in type 1 (or subgroup D in the classification of Lyness et al.) patients have a diffuse loss of rod sensitivity with patchy and later loss of cone sensitivity while in type 2 (or subgroup R in the classification of Lyness et al.) patients have regionalized and a combined loss of rod and cone sensitivity.

Birch and Fish[2] recorded rod and cone responses with scotopically matched blue and red stimulus lights, respectively, so that both amplitudes were equal in normal eyes. They calculated the rod/cone amplitude ratio (R/C ratio) in 35 patients with widespread retinal degeneration. They arbitrarily defined cone-rod degeneration as an R/C ratio greater than 1.0 and the typical rod-cone form of RP as an R/C ra-

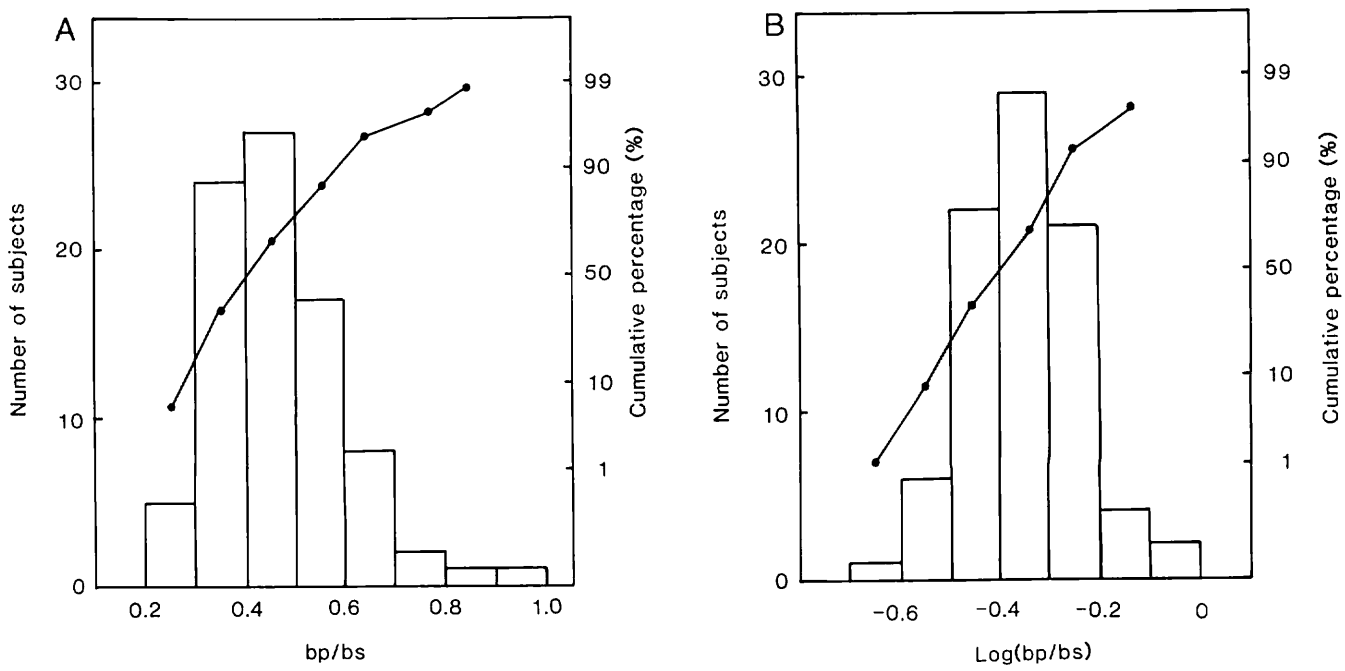

FIG 65–1.
Histograms and their cumulative percent frequencies on a probability paper for bp/bs **(A)** and log (bp/bs) **(B)** of 85 normal subjects (38 males and 47 females, aged 3 to 77 years). The histogram of bp/bs is asymmetrical, with its peak deviating toward the lower end and the line of its cumulative percent frequency curving up in the middle, while the histogram of log (bp/bs) appears almost symmetrical, and the line is straight, which indicates that bp/bs follows the log normal distribution.[4]

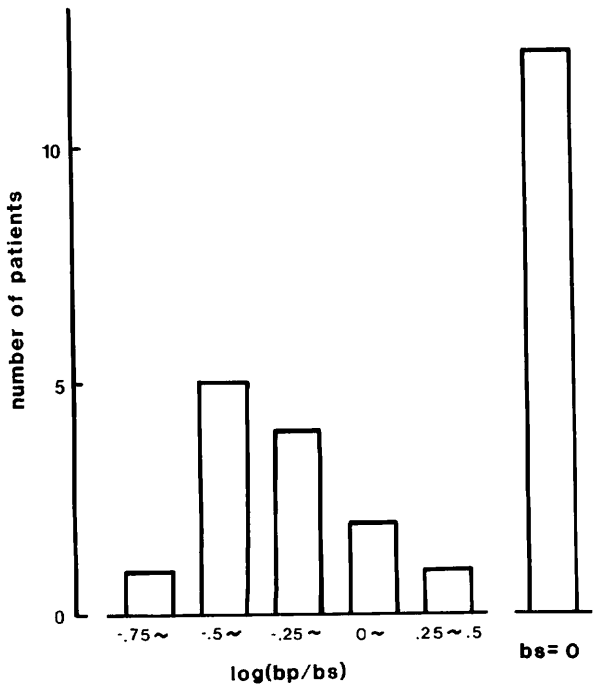

FIG 65–2.
Histogram of log (bp/bs) in patients with autosomal dominant RP and photopic ERGs larger than 20 μV. The *rightmost bar* represents patients with nonmeasurable scotopic ERGs (i.e., less than 5 μV) whose log (bp/bs) values were considered to be more than 0.5. The remaining bars seemed to form a single group with its distribution peak between −0.5 and −0.25.

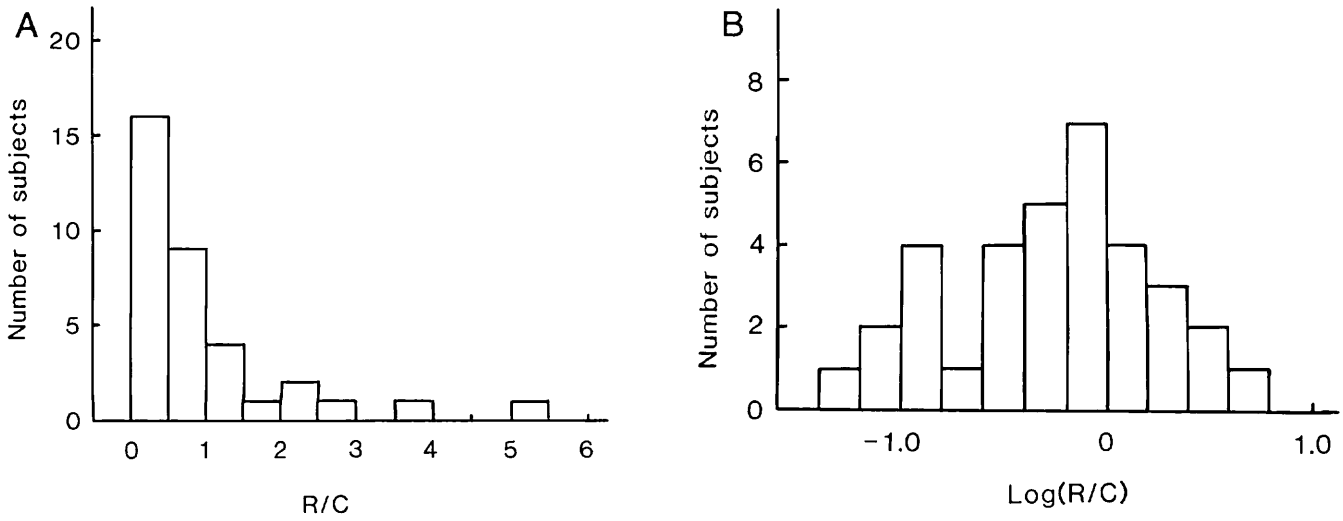

FIG 65–3.
Histograms of R/C (i.e., the ratio between rod and cone ERG amplitudes elicited by scotopically matched blue and red stimulus lights) **(A)** and log (R/C) **(B)** in patients with diffuse photoreceptor disease who appeared in the literature.[2] While the author defined cone-rod degeneration as an R/C greater than 1.0 and RP as less than 1.0, the distribution pattern of log (R/C) is suggestive and indicates that two groups can be divided around −0.8 of log (R/C).

tio less than 1.0. However, the distribution pattern of the log of the ratio in all of their subjects indicated that two groups can be divided by the borderline of −0.8 rather than 0 (log 1.0) (Fig 65–3). While their subjects included patients with autosomal recessive inheritance and isolate cases as well, the patients with log (R/C) less than −0.8 probably conform to our type 1 patients with autosomal dominant RP, and the patients with log (R/C) between −0.8 and 0.8 conform to our type 2 patients.

Evaluation of rod and cone function by log (bp/bs) or log (R/C) is a useful classification for patients with RP. It is expected that it will also provide a diagnostic and classifying methodology for other photoreceptor diseases such as choroideremia, cone dystrophy, and RP syndromes with systemic involvement.

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