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Dark adapted cone/rod thresholds and ERGs in Duchenne muscular dystrophy

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Footnotes

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Abstract

Purpose : An electronegative ERG (b-wave < a-wave amplitude) is present in patients with Duchenne muscular dystrophy (DMD) with genetic alterations downstream of exon 30 because of the absence of dystrophin proteins (Dp427 and Dp260) in the outer plexiform layer. Here we examined if this retinal change affects dark adaptation.

Methods : Participants were 17 DMD patients (13.5±4.3 years old) and 34 age-similar controls (16.9±6.6 years old) with normal ophthalmological exams. Dark adaptation measurements were applied to the dominant eye using a dark adaptometer (Roland Consult). Detection thresholds to flashes of red (625 nm; cone function) and green (527 nm; rod function) LEDs (2° diameter) were measured after one minute of bleaching (3000 cd/m^2). Initially, 8-minutes of interleaved red and green thresholds were measured. After additional 5-minutes of dark adaptation, a second set of threshold measurements to green stimuli only was performed during the subsequent 6-minutes. Cone thresholds were obtained by averaging the luminances required to detect the red stimulus during the last minute of the first protocol. Rod dark adapted thresholds were obtained by averaging the luminance levels at the three last minutes of the second protocol (Figure 1). Dark-adapted ERGs were recorded to 10 cd.s/m^2 white light flashes and individual b-/a-wave amplitude ratios were obtained. Comparisons were performed with ANOVA and Spearman coefficient correlations.

Results : Mean cone threshold (log cd/m^2) did not differ statistically (p=0.198) between controls (mean=-1.78±0.38) and DMD patients (mean=-1.67±0.71). Final rod threshold was disturbed for patients with genetic alterations downstream of exon 30 (mean=-3.13±0.64; p<0.001) relative to controls (mean=-4.23±0.27). No statistical difference was found between controls and DMD patients with alteration upstream of exon 30 (mean=-4.00±0.49; p=0.073). Mean ERG b/a ratios were significantly higher (p<0.001) for controls (mean=1.47±0.22) compared to upstream 30 (mean=1.18±0.32) and downstream 30 DMD patients (mean=0.65±0.37). ERG b/a ratios were negatively correlated with dark adapted rod thresholds (Spearman=-0.618; p=0.005).

Conclusions : The larger the magnitude of ERG alteration the greater the loss in rod dark adapted sensitivity. Dystrophin product Dp260, which is required for normal dark adapted ERG, also plays a role in rod sensitivity of the dark-adapted human retina.

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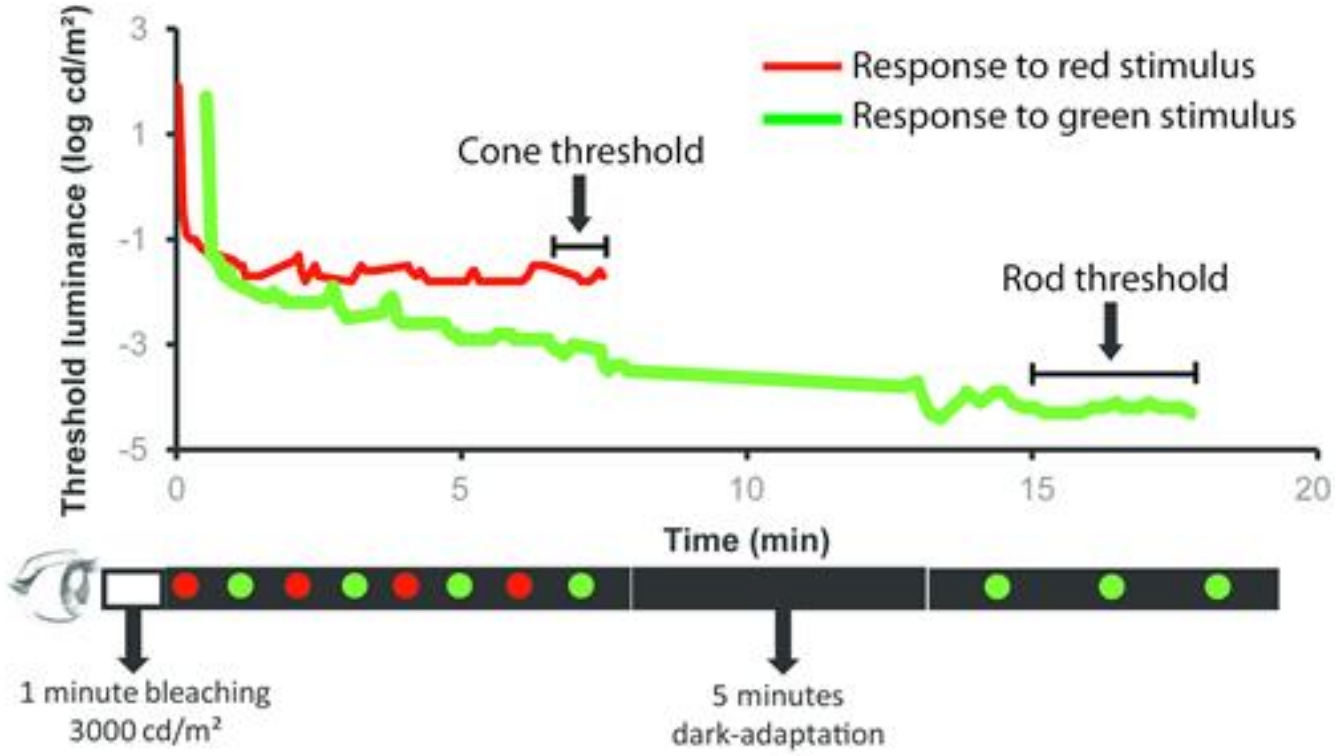

Figure 1. Dark adaptation curves showing threshold luminance as a function of time in the dark.

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