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Color vision deficiency in retinitis pigmentosa

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Abstract. *Purpose:* This study examined which color vision test was superior in the detection of acquired color vision deficiencies and the relative magnitudes of red–green (RG) and blue–yellow (BY) discrimination losses in retinitis pigmentosa (RP). *Methods:* Color vision examinations were conducted using Standard Pseudoisochromatic Plates Part 2 (SPP Part 2) and a flicker threshold test (TMT) in 21 RP and 21 age- and sex-matched normals. The TMT test consists of luminance, BY and RG flicker tests. Relative Operating Characteristic analysis (ROC) was used to determine the effectiveness of each test. Bigger ROC area gain and high performance (%) indicate a better test. Regression and correlation analyses were used to determine the relative magnitudes of RG and BY discrimination losses for the SPP Part 2. For TMT the findings were represented graphically where the mean log sensitivity was plotted against the frequency of mean luminance; RG and BY flicker individually and the range was plotted with the 5th and 95th percentile. *Results:* Both tests were able to determine BY and RG acquired color vision deficiencies in RP patients. SPP Part 2 showed the highest area gain (0.39) and performance (78%). The TMT generally showed less area gain and performance compared to SPP Part 2. Pearson's correlation coefficient analysis on the RG and BY SPP Part 2 test plates showed a strong relationship ($r=0.96$, $p<0.0001$) between RG and BY discrimination losses suggesting that RP patients suffer roughly equal losses in RG and BY discrimination. Analysis of the TMT also showed an equal loss of sensitivity across all the frequencies in all three tests suggesting an overall reduction of sensitivity confirming the non-specific frequency and non-color specific loss. *Conclusions:* The SPP Part 2 appears to be a superior test compared to TMT in detecting acquired color vision deficiencies in RP patients. RP subjects also suffer both RG and BY acquired color vision deficiencies.

Keywords: Color vision; Retinitis pigmentosa

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1. Introduction

Color vision is well known to be affected in retinitis pigmentosa (RP) [1–4]. However, identifying the type of acquired color vision loss in RP may not be straightforward. Clinical color vision tests are simplified versions of psychophysical methods and mainly based on pigment colors. This makes them relatively inexpensive and requires little, if any, calibration. Although much research has been devoted to color vision in RP [2–5], little information is available on the specificity and sensitivity of the SPP Part 2 test in detecting color vision losses in RP subjects. The flicker threshold test named temporal modulation threshold technique (TMT) is a psychophysical means of determining color vision loss. Unlike congenital color deficiencies, acquired deficiencies may involve dysfunction of the cones or post-receptoral neurones or both. Therefore, psychophysical testing can aid in evaluating the site of an acquired deficiency [6,7]. This study examined (1) the effectiveness of the Standard Pseudoisochromatic Plates Part 2 (SPP Part 2) and the flicker test using the temporal modulation threshold method (TMT) in identifying color vision deficiencies in RP are assessed. (2) The relative magnitudes of red–green (RG) and blue–yellow (BY) discrimination losses in RP were compared.

2. Method

Color vision examinations were conducted using SPP Part 2 and a flicker threshold test (TMT) in 21 RP and 21 age- and sex-matched normals.

2.1. Testing of the SPP Part 2 and the flicker threshold test (TMT)

The illumination used for SPP Part 2 was provided by Crompton coolwhite daylight fluorescent tubes, which were rated DE when assessed in accordance with the procedures of the CIE method of assessing conformance with daylight simulators CIE (1981) [8]. No restriction upon holding the test plates or time limits were imposed upon subjects during testing [4]. The color vision test was conducted monocularly, with the better eye and with any appropriate near correction that was routinely used by the subject. The subjects are required to identify the digits on each plate for the SPP Part 2 test. A flicker threshold test was conducted using the temporal modulation threshold technique (TMT). This method comprises of a Maxwellian view produced from three light emitting diodes (red, yellow-green and blue). These sources are modulated sinusoidally and may be run in phase (luminance flicker) or in counter phase (RG and BY flicker). The amplitude and/or frequency of the flickers can be controlled. The waveform can be selected (sine, square, ramp up, ramp down).

3. Results

3.1. Effectiveness of tests in detecting color vision deficiency in RP

The effectiveness of tests in detecting color vision deficiency in RP patients was determined. The findings showed that both the SPP Part 2 and the TMT tests were able to determine blue–yellow (BY), and red–green (RG) acquired color vision deficiencies in RPs. The performance of the subjects with the SPP Part 2 (as a whole test, BY, RG) and the TMT tests (luminance flicker, BY and RG Flickers) were further analyzed using the ROC where the specificity and sensitivity was calculated.

Table 1
Summary of the ROC area gain and performance of the 2 tests

Tests		ROC area gain	Performances (%)
Pseudoisochromatic test plates	SPP Part 2	0.39	78
	SPP Part 2 blue–yellow	0.39	78
	SPP Part 2 red–green	0.31	62
Flicker test	Luminance flicker (5 Hz)	0.30	60
(Temporal modulation tests)	Blue–Yellow flicker (5 Hz)	0.27	54
	Red–Green flicker (5 Hz)	0.28	56

The ROC areas and percentage of performances were summarized in Table 1. SPP Part 2 showed the highest area gain (0.39) and performance (78%). The TMT generally showed less area gain and performance compared to SPP Part 2.

3.2. Comparison of the relative magnitudes of red–green (RG) and blue–yellow (BY) discrimination losses in RP

To determine the relative magnitudes of RG and BY discrimination losses in RPs, correlations and regression analyses were conducted for SPP Part 2. The percentage of errors in SPP Part 2 RG plates was plotted against the percentage of errors in SPP Part 2 BY (Fig. 1). Analysis of Pearson's correlation coefficient showed that in the graph, there was a strong relationship between the loss of BY and RG in RP subjects ($r = 0.96$ $p < 0.0001$). These findings suggest that the subjects with RP were making equal proportions of errors in the SPP Part 2 BY plates and SPP Part 2 RG plates. For TMT Flicker tests, the range for normal was plotted with the 5th and 95th percentile. The results of the TMT tests showed that there was an overall reduction of sensitivity in the mean luminance, RG and BY flicker test results across all frequencies (Fig. 2). The mean color flicker performances however were within normal limits. These findings showed that the magnitude of the overall reduction was very similar across the range of frequencies tested. This indicates that there was a non-frequency specific and a non-color specific loss of color vision in RP subjects.

4. Discussions

A perfect test will show a specificity and sensitivity of one with the ROC area being 0.50; percentage of performance (%P) of the test is 100%. Clinically, this can be interpreted as larger ROC area, higher level of accuracy in separating normals from the

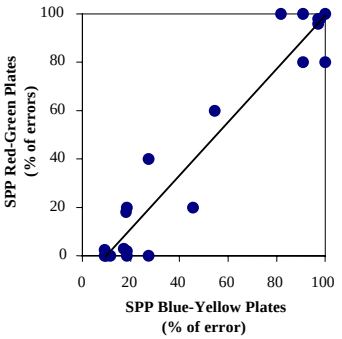

Fig. 1. Percentage of error in SPP red–green against SPP blue–yellow $r = 0.96$ $p < 0.0001$.

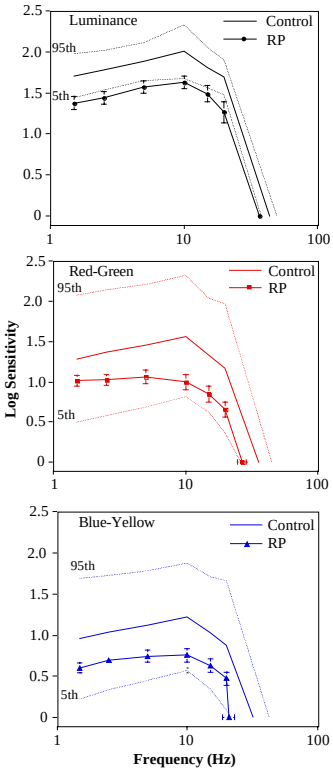

Fig. 2. Mean luminance, red–green, blue–yellow flickers for RP and control for TMT test.

color vision defectives. In this study, ROC area for SPP Part 2 as a whole test was 0.39 and % P was 78%. The BY plates in the SSP Part 2 showed a similar percentage of performance with the SPP as the whole test followed by the RG plates. These results are in agreement with previous studies suggesting that the SPP Part 2 able to identify acquired BY and RG deficiency [4,9–11]. On the other hand, other researchers have found that the color vision losses in RPs could be either BY or RG deficiencies [1,2,4]. One possible explanation is that the color vision tests used in those experiments were not specifically designed to detect acquired color vision loss. Other possible explanation could be due to the disease stage and the degree of macula involvement in the selected test subjects. Even though the population of RP subjects in this study is limited, it can be concluded with certainty that the SPP Part 2 is an effective BY (tritan) and RG screening tool in RP subjects.

This study also attempted to evaluate a flicker test (luminance and color flicker) to identify if this test is superior in the detection of the acquired color vision deficiencies in RP. The findings showed that there was an equal overall reduction in the flicker sensitivity for all frequencies and in luminance, BY and RG flicker in RPs. The findings indicate that any one frequency, any luminance, or color could be used to measure visual sensitivity losses. ROC curves analysis indicate that luminance flicker showed the biggest area of separation indicating that luminance flicker test is a more sensitive test in detecting losses

of cone function in RPs. These results are in substantial agreement with that of previous researchers [12–14]. The interpretations of psychophysical measurements in acquired color deficiencies of RP subjects must be interpreted with care. This is because it may be influenced by a number of complicating factors. These include the possibility of congenital red–green deficiencies, which affects about 8% of the male population [12] and the effects of macular and lens changes.

5. Conclusions

From the analysis of the SPP Part 2 test and the TMT tests, it can be concluded that both tests have the capability of determining both BY and RG deficiencies. The SPP Part 2 appears to be a superior test compared to TMT in detecting acquired color vision deficiencies in RP patients In the TMT analysis, it can be concluded that there was an equal loss of sensitivity across all the frequencies in luminance, BY and RG flicker. These findings demonstrate that there was an overall reduction of sensitivity of luminance and color flicker confirming that there was a non-specific frequency and a non-color specific loss in RP. It can then be concluded that RP subjects suffer both RG and BY acquired color vision deficiencies that become more evident as the disease progresses.

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