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# Principles and Practice of Clinical Electrophysiology of Vision

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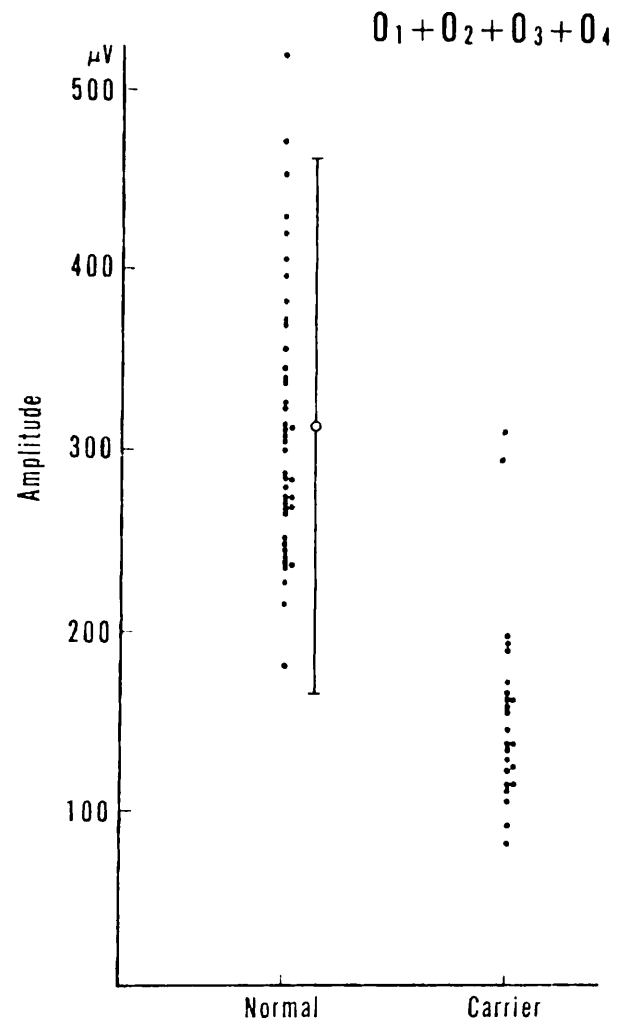
# Carrier State of Congenital Stationary Night Blindness

Yozo Miyake

Only a few studies<sup>1, 2, 4</sup> have been performed on the visual functions of the female carrier in X-linked recessive congenital stationary night blindness (CSNB) with myopia (complete type in our classification).<sup>3</sup> Patients with this type of CSNB have an absent rod electroretinogram (ERG), a good cone ERG, and a Schubert-Bornschein-type ERG with nearly absent oscillatory potentials.

We found significant abnormalities of oscillatory potentials in female carriers in X-linked recessive CSNB<sup>2</sup> (complete type<sup>3</sup>). Figure 95-1 shows the distribution of the amplitude of oscillatory potentials ( $O_1 + O_2 + O_3 + O_4$ ) in 12 female carriers in comparison with that of 42 age-matched normal controls. The ERG was recorded with a single bright white flash in the dark after 30 minutes<sup>1</sup> dark adaptation. The amplitude of the oscillatory potentials in carriers was significantly smaller than in normal eyes. The peak time of oscillatory potentials and the amplitudes of a-, b-, rod b-, and cone b-waves were within the normal range.

Young et al.<sup>4</sup> confirmed that the amplitude of oscillatory potentials is reduced in carriers. They recorded the oscillatory potentials under four stimulus conditions and analyzed them in time and frequency domains. The results showed that the best condition



**FIG 95-1.**

Variation of the amplitudes of oscillatory potentials ( $O_1 + O_2 + O_3 + O_4$ ) in normal subjects and carriers of X-linked recessive CSNB (complete type).

for discriminating between carriers and normal subjects was when the flash was blue and the eye was dark-adapted. In the frequency domain, optimal discrimination occurred when examining the power content of the oscillatory potentials at a center frequency of about 130 Hz with a 70-Hz-bandwidth window. In the time domain, optimal discrimination occurs when examining the amplitude of the third peak of the response.

Kewish et al.<sup>1</sup> performed dynamic ERG to differentiate among males with X-linked recessive CSNB with myopia and nystagmus, obligate carriers, and normal subjects. They evaluated the amplitude of ERG a-, cone b-, and rod b-waves and oscillatory potentials. They concluded that oscillatory potentials alone were enough to enable discrimination of the three groups.

These previous results in studying the carriers in X-linked recessive CSNB with myopia (complete type)<sup>3</sup> indicated that the oscillatory potentials are

key in discriminating between normal and female carriers.

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