SECTOR RETINITIS PIGMENTOSA

ELECTROPHYSIOLOGICAL AND PSYCHOPHYSICAL
STUDY OF THE VISUAL SYSTEM

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ABSTRACT

Combined clinical, psychophysical and electrophysiological examinations of the visual function were performed in two patients affected by sector retinitis pigmentosa. Psychophysical dark adaptation measurements, the electroretinogram (ERG) and the electro-oculogram (EOG) revealed that a larger area of the neural retina and pigment epithelium were pathologically involved than revealed by ophthalmoscopy and visual field. Five years of observation showed a stationary retinal defect in one patient, while in the other a slight clinical but a marked electroretinographical deterioration was found over a period of twelve years. However, visual acuity and the visually evoked potential (VEP) remained normal. In spite of subnormal ERG amplitudes, the photopic and scotopic peak latencies were normal in both cases. In the ophthalmoscopically normal relatives of one patient slight dark adaptation impairments, as well as EOG and ERG disturbances, were detected.

INTRODUCTION

Sector retinitis pigmentosa is a rare type of tapetoretinal dystrophy characterized by a sectorial retinal affection which is almost symmetrical in both eyes and exhibits with the ophthalmoscope pigment clumps and vascular changes typical for retinitis pigmentosa.

Following the first case published by Bietti in 1937, eighty-three affected patients have been reviewed up to 1970 (Bisantis, 1971) and since then only few additional cases have been dealt with (Pasco, 1970; Franck et al., 1971; Berson & Howard, 1971; Ivandić, 1972; Thaler et al., 1973; Hellner & Rickers, 1973). Autosomal dominant inheritance was the common hereditary pattern, while the autosomal recessive and sex-linked traits were less frequently reported. In contrast to most types of retinitis pigmentosa, the sector type is considered stationary or only very slowly progressive, and the fovea preserves its normal function even at old age. Usually the retinal lesion is localized in both eyes symmetrically in the nasal quadrants, with bilateral scotomata, related, more or less, to the ophthalmoscopically visible retinal defect. Using fluorescein angiography Krill et al., (1970) showed that the pigment epithelium disturbance extend beyond the sectorial area into ophthalmoscopically normal retina. In keeping with the affected pigment
epithelium, the resting potential measured by the electro-oculogram (EOG) revealed subnormal values in all examined cases (Graham, 1963; Krill et al., 1970; Thaler et al., 1973; Hellner & Rickers, 1973).

Studies of retinal function by means of electroretinogram (ERG) showed in almost all sector retinitis pigmentosa patients subnormal amplitudes (Lisch, 1955; Küper, 1960; Graham, 1963; Franceschetti et al., 1963; Ponte, 1966; Hommer & Wohlzogen, 1970; Krill et al., 1970; Bisantis, 1971; Franck et al., 1971; Berson & Howard, 1971; Ivandić, 1972; Thaler et al., 1973; Hellner & Rickers, 1973) and almost extinct responses in two older patients (Vukovich, 1959; Haase & Hellner, 1965). In infants the subnormal ERG precedes the appearance of the sectorial retinal lesion (Hellner & Rickers, 1973). Follow-up examinations with the ERG did not disclose changes in amplitude during four (Krill et al., 1970), seven (Hellner & Rickers, 1973) and even ten years (Hommer & Wohlzogen, 1970). Clinically normal adult members from two families of affected individuals were examined by the ERG and revealed normal responses (Haase & Hellner, 1965; Berson & Howard, 1971). Regarding the temporal aspects of the ERG normal cone and rod implicit times (peak latencies) were found in sector retinitis pigmentosa patients (Berson & Howard, 1971), in contrast to delayed implicit time found in widespread retinitis pigmentosa (Berson et al., 1969; Berson & Kanters, 1970) and Hunter disease (Abrham et al., 1974). The disadvantage of most reports dealing with the ERG is that nonstandardized and, especially in the older studies, less sophisticated methods than possible now were employed.

While psychophysical measurements of dark adaptation yielded normal results in some of the cases (Ponte, 1966; Thaler et al., 1973), obvious impairment of the scotopic function of the retina was usually found (Bisantis, 1971). With dark adaptation perimetry Krill et al. (1970) found elevated thresholds not only in the ophthalmoscopically affected quadrants, but also all over the retina. On the other hand, Berson & Howard (1971) have found scotopic impairment only in the disturbed quadrants. It is, therefore, questionable whether the ophthalmoscopically normal retina around the sectorial defect is really healthy. In addition, findings like subnormal ERG with normal implicit time, subnormal EOG's, slowly deteriorating visual fields but preserved central vision, make the study of sector retinitis pigmentosa worthy of further investigations.

The present study deals with two patients with sector retinitis pigmentosa who underwent clinical examinations, ERG, EOG, VEP recordings, and psychophysical measurements of dark adaptation. The disease was followed over a period of five years in one patient and of twelve years in the other. In order to trace possible retinal disturbance in the clinically normal retina,