Does Unilateral Retinitis Pigmentosa Really Exist?

An ERG and EOG Study of the Fellow Eye

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Diagnosis of unilateral tapetoretinal dystrophy (retinitis pigmentosa) is certified only if the case meets the following requirements (Franceschetti, Francois and Babel, 1963—Report of the French Ophthalmological Society): 1. Exclusion of inflammatory origin of the affection; 2. Absence in the fellow eye of symptoms of tapetoretinal dystrophy and presence of a normal ERG.

The latter condition is said to allow an early and certified diagnosis. If no ERG is available, a sufficiently long observation period is compulsory, fixed tentatively, by Francois, at 5 years.

Publication of 4 new cases—of which 3 undoubtedly fit the above criteria, but in which the standing potential of the fellow eye demonstrated over a 1–2 years' period a progressive deterioration—makes clear that the list of requirements allowing the earliest possible diagnosis needs to be completed at least with the demand for a normal standing potential of the fellow eye. But even this requirement does not guarantee the stationary condition of the fellow eye.

A long observation period is still inevitable before one can make the diagnosis with reasonable certainty.

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Discussion

Schweitzer: In the EOG curves which you showed us the negative phase, which one expects after about 12 minutes of dark adaptation, was absent. Is not this abnormal?

Henkes: As the result of our experimental conditions—the patients are partially adapted to the dark during the application of the electrodes—marked changes in the resting potential at the end of the following period of 12 minutes of complete adaptation to the dark do not occur.

Winkelman: Prof. Henkes has mentioned a patient of Dr. Jager's and mine. I could let him see another patient with an unquestionable unilateral retinitis pigmentosa; the diagnosis here is not open to doubt: pigmentations, ring scotoma, unilateral abnormal adaptation curve, unilateral absent ERG while the other is normal; the EOG has not been determined. I have known this patient for more than 10 years and this state of affairs has remained unchanged. Does Prof. Henkes not think that his standard for the EOG is rather arbitrary? In my opinion, genuine unilateral retinitis pigmentosa is extremely rare, but does occur.

Henkes: Every standard which is laid down is arbitrary in a sense. I do not so much deny the existence of genuine unilateral tapeto-retinal degenerations, but I do contest—on the grounds of, in my opinion, a reliable criterion, i.e. the presence of a “normal” EOG—the relatively frequent occurrence of this condition as would appear from the literature.

Mrs. Komproe: One can try to explain the unilateral occurrence of the tapetoretinal degeneration by a developmental difference between the two eyes. Was this also expressed by a difference in refraction?

Henkes replies that no obvious anisometropia was present.

Mrs. Schappert: Franceschetti, Francois and Babel are of the opinion that the diagnosis can be made from the result of the ERG without a longer period of observation. I consider that this conclusion is not justified and that the five-year observation period must be maintained.

Schweitzer asks for the geneticist's opinion about unilateral tapeto-retinal degeneration.

Waardenburg: No unilateral case has so far been found in association with one or more bilateral cases in a family following any of the three types of heredity. Landolt's case (1959) of unilateral occurrence in a father and child is probably due to reversal of the dominance in heterozygotes.