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VISUAL FIELDS IN RETINITIS PIGMENTOSA

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Abstract

Retinitis Pigmentosa is an hereditary retinal dystrophy which is characterised by night blindness and visual field defects.

This paper describes the visual fields of twenty consecutive patients attending the Ocular Diagnostic Clinic of the Royal Victorian Eye and Ear Hospital. The fields have been classified on a scale from one to seven. The patients with the most severe field defects also tended to have the highest dark adaptation thresholds and extinguished electroretinograms. Those patients with the earliest age of onset of symptoms had the most severe field defects.

Partial ring scotomata showed sparing in the nasal or lower region, and when annular scotomata broke through to the periphery they did so in the upper part of the field.

It is anticipated that some types of Retinitis Pigmentosa may be treatable within ten to twenty years so it is important to identify patients and to gather data.

Key words: *Retinitis Pigmentosa, annular scotoma, dark adaptation, Goldmann perimetry, visual field, hereditary retinal dystrophy.*

Retinitis Pigmentosa (RP) can be defined as an hereditary retinal dystrophy which diffusely affects photoreceptors and pigment epithelium. It is characterised by its onset, (usually in adolescence), by night blindness, and by constriction of the visual fields.

Features include signs of retinal pigmentary changes, constriction of arterioles and optic atrophy. There is a steady progression of visual loss over the years, with macular problems usually developing at a later stage.

According to Krill[1] the classical field defect is an annular scotoma, which is nearly always present in early cases, located between 30° and 50° of fixation. It may not be a complete ring. It later spreads both towards the centre and peripherally, in severe cases leaving a temporal and central island of field, which is insufficient for orientation. The patient can be legally blind as a result of field loss, whilst still retaining reasonably good central vision. Ultimately the remaining field can be lost, the centre being the last to go. Generally, the earlier the presentation, the worse the prognosis, as in late onset RP the field loss is not so great.

It has been stated[2] that the prevalence of RP in Victoria may be as high as one in two thousand. Bunker et al[3] found a prevalence of one in four thousand seven hundred in Maine, U.S.A.

There are three different modes of inheritance:

(i) Autosomal recessive, which is the most common. An example of this group is Usher's syndrome, which consists of sensorineural deafness (usually congenital) and RP with onset in the teenage years.

(ii) Autosomal dominant, which is less common

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(iii) X linked recessive, which is the most severe, and the least common.

The aim of this paper is to introduce a classification system for the visual fields of RP patients, and to describe data on twenty patients with this condition.

SUBJECTS

A consecutive sample ($n = 20$) was taken over a three month period (3.4.86 to 3.7.86) of patients attending the Ocular Diagnostic Clinic of the Royal Victorian Eye and Ear Hospital, who had a definite diagnosis of RP and who were able to give a reliable visual field. Two eyes were excluded from the series due to additional pathologies.

The subjects' ages ranged from nine to 68 years, with a mean age of 43. There were 13 males and seven females. The age of onset ranged from less than 5 years to 56 years, with a mean age of 18.

Nine were diagnosed as definite and four as probable autosomal recessive type (with five cases of Ushers syndrome and one congenital amaurosis of Leber). There were four definite and three probable cases of autosomal dominant type. None were of the X linked recessive type.

Forty-five per cent of the eyes had visual acuity of 6/9 or better, 13% had 6/60 or worse.

All patients had a full ophthalmic examination including fundus examination, visual acuity, visual fields, electroretinography, dark adaptometry, and colour vision testing (using the Ishihara test).

The visual fields were tested using the Goldmann perimeter, usually using targets size I intensity 4, size III intensity 4 and size IV intensity 4, as recommended by Marmor et al.[4] The standard method was used with movement of the target from the periphery to the centre, and from non seeing to seeing areas.

Dark adaptometry was performed using the Goldmann/Weekers dark adaptometer, with the patient sitting in a totally dark room whilst their threshold sensitivity to light was measured at specific intervals.

RESULTS

To aid in the comparison and description of patients, the following classification devised by

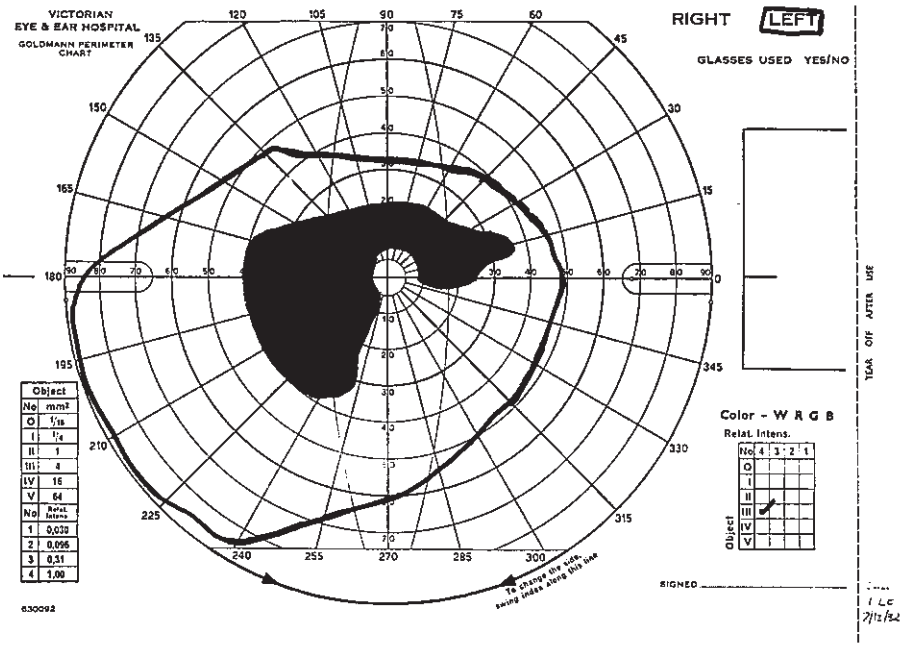

Figure 1: Example of Field Type 2 (partial ring scotoma).

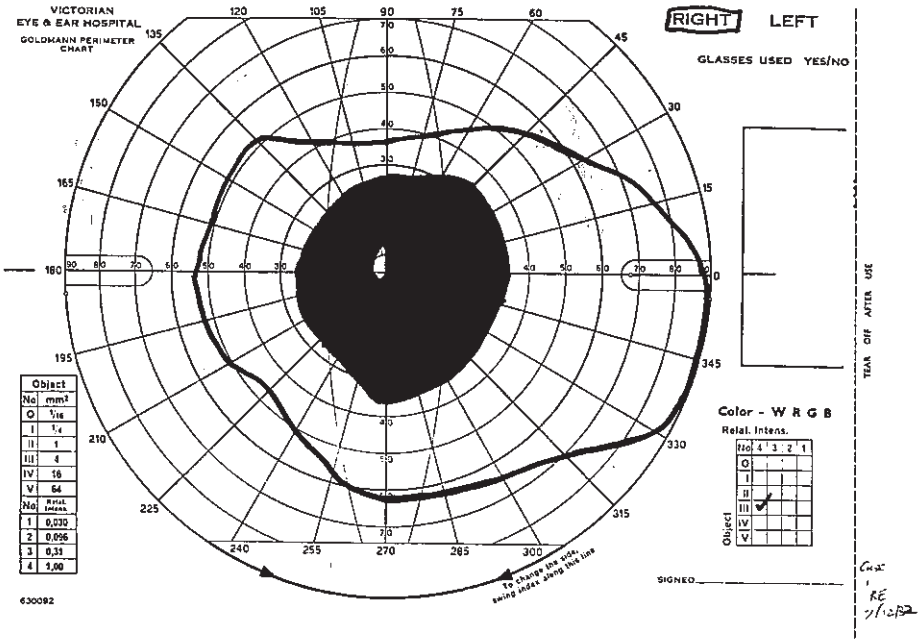

Figure 2: Example of Field Type 3 (complete ring scotoma).

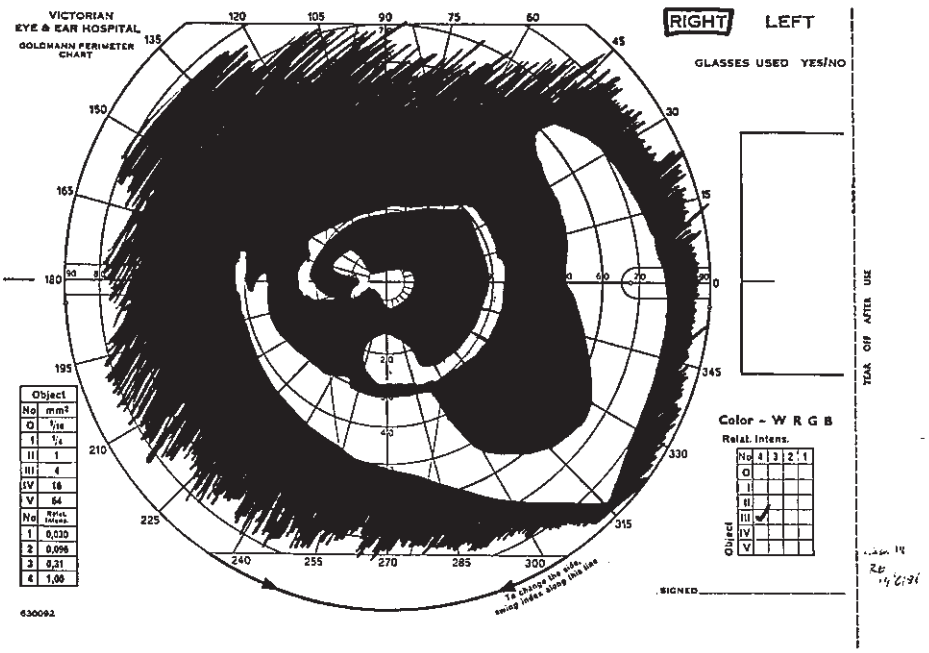

Figure 3: Example of Field Type 4 (ring scotoma with partial breakthrough to the periphery).

Dr H. MacLean of the Ocular Diagnostic Clinic was used. This classification uses the Goldmann size III intensity 4 target wherever possible. If a larger target is needed, the smallest possible is used.

Type 1. Full field.
Type 2. Partial ring scotoma. (Fig. 1.)
Type 3. Complete ring scotoma. (Fig. 2.)
Type 4. Ring scotoma with partial breakthrough to the periphery. (Fig. 3.)
Type 5. Complete breakthrough to the periphery, but with a radius of more than 30° in one or more sectors. (There were no eyes in this category in this series.)
Type 6. Constricted to a radius of less than 30° of fixation. (Fig. 4.)
Type 7. Constricted to a radius of less than 10° of fixation. (Fig. 5.)

Table 1 shows the incidence of patients in each of these categories.

TABLE 1
Showing Incidence of Each Field Type of the 38 Eyes Studied

Field type	1	2	3	4	5	6	7
Number in series	2	4	3	7	0	7	15

Electroretinography (ERG)

The ERG is a mass retinal response which is only affected if a large proportion of the retina is diseased or malfunctioning. None of the eyes in this study had a normal ERG, and, as would be expected, the eyes with the most severe field defects were more likely to have extinguished ERGs. The ERG was extinguished in 13 out of the 15 eyes with type 7 fields, six out of the 14 eyes with type 4 or 6 fields, and in only two out of the nine eyes with types 1 to 3 fields.

Dark Adaptation

A moderate correlation ($\rho + 0.46$) was shown to exist between the type of field and the dark adaptation threshold. Only one patient with type 1 fields and one with type 2 fields had dark adaptation thresholds within normal limits.

Onset

As the exact age of onset is often difficult to determine, for the purposes of this study it was regarded as five years if an adult stated that symptoms had been present all his life. If the age of later onset was unknown, but the adult stated that it had been present from childhood, then the onset was recorded as 10 years.

Comparison of 11 cases with onset before 15 years with five cases of onset after 30 years shows that those with early onset had a slightly greater proportion of extinguished ERGs, slightly higher dark adaptation thresholds, and poorer vision. Thirty-three per cent of early onset eyes had vision of 6/9 or better, whereas this vision was obtained in 67% of the late onset eyes. The cases with the earliest onset also tended to have the most severe field loss.

Duration

Although there was no particular relationship between severity of field defect and age, generally the longer the duration of the condition the more severe is the field defect. This becomes particularly apparent for those with onset after 30 years, when there is a strong correlation ($\rho + 0.83$). Some of the early onset cases reached type 6 or 7 fields in less than 10 years.

Residual Central Field

There were 11 eyes of eight patients with a central field of less than 5° radius, although in five of these some peripheral field could be demonstrated using a larger target. Patients in this group tended to be older, with a longer duration of symptoms, and visual acuity and colour vision were also worse than the group as a whole. There were also, proportionally, more males in this group.

Genetic Type

Of the 25 eyes of patients with autosomal recessive type, 17 (66%) had type 6 or 7 visual fields, whereas of the autosomal dominant type (13 eyes) only 5 (40%) had severe field loss. These two groups were of similar ages, and had similar ages of onset. ERGs and dark adaptation thresholds were also similar for the two groups.

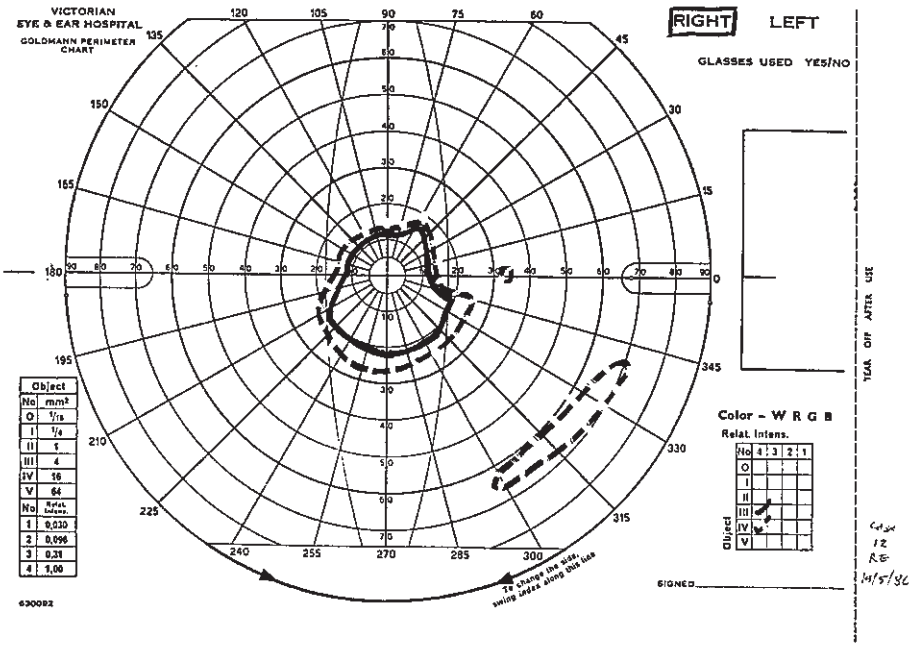

Figure 4: Example of Field Type 6 (field constricted to a radius of less than 30 of fixation).

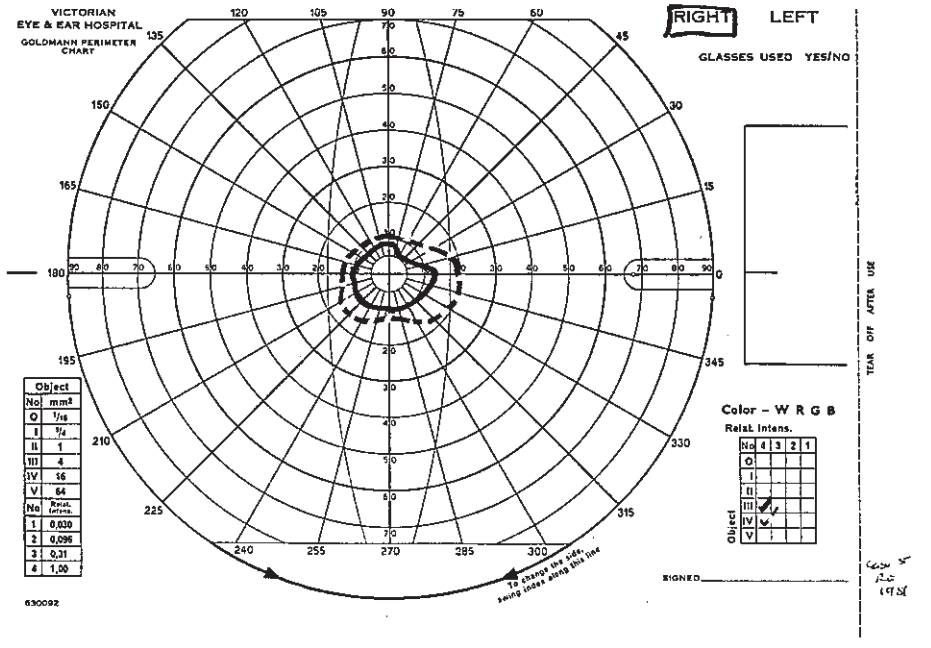

Figure 5: Example of Field Type 7 (field constricted to a radius of less than 10 of fixation).

DISCUSSION

In this series of thirty-eight eyes all cases of partial ring scotoma (type 2) had sparing of the nasal or lower nasal region; in all cases of annular scotoma with partial breakthrough to the periphery (type 4) the breakthrough was in the upper or upper and temporal part of the field.

Krill[1] states that the ring scotoma in RP usually occurs between 30° and 50° of fixation. In the present study it was not possible to specify the precise position of the initial field defect, because most were too advanced. At the time of this study seven eyes (of four patients) had partial or complete ring scotoma (type 2 or 3). Examination of the earlier fields of the five patients who had attended previously, provided a further five eyes (of three patients) with fields type two or three. So 12 eyes of seven patients are considered.

In these 12 eyes the inner boundary of the scotoma occurred between 2° and 5° in seven eyes, at 10° in three, at 15° in one and at 30° in one. The outer boundary occurred between 10° and 20° in four eyes, between 30° and 50° in seven, and at 70° in one. So seven out of 12 eyes had a scotoma with the outer boundary between 30° and 50°, and one more peripheral, but in all but one eye the inner boundary was nearer to fixation than 30°. This finding was surprising, it would be interesting to study patients at an earlier stage of the disease to discover whether most scotomata do start between 30° and 50° and extend centrally at a greater rate than peripherally.

According to Krill[1] a temporal island of field often remains as well as the centre. In this study, in all eyes which had small peripheral islands of field, these were located in temporal, lower temporal or lower region at about 70° from fixation and were usually approximately 10° wide.

All eyes had an area of field remaining in the centre. If the twenty-six eyes with the worst field defects are considered, 15 had only a central island of vision, four had a temporal arc of field between 60° and 80°, three had a lower temporal arc between 70° and 90° and four had a lower arc between 30° and 55°.

Although conditions such as congenital stationary night blindness, vitamin A deficiency and night myopia can cause difficulties with night vision, perimetrists should be alterted to the possibility that a patient with this symptom may have RP, and if so, peripheral fields are required. The upper, temporal and lower areas should be searched for scotomata between 15° and 50°.

Visual field testing is an important part of the examination of RP patients, and provides information to aid diagnosis and assessment of progress of the disease, and for assessment of eligibility for the blind pension. In addition, early diagnosis provides the opportunity for genetic and career counselling.

As a result of genetic research[5] it is possible that some forms of RP may be treatable within twenty years. With the possibility of treatment within the lifetime of some of the current patients, or their children, it is becoming increasingly important to maintain comprehensive records of these patients.

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References

1. Krill AE, Archer D. Krill's hereditary retinal and choroidal diseases. Vol II, Hagerstown, Harper and Row, 1977.
2. Dickinson P. Classification of retinitis pigmentosa (paper given at RACO Victorian Branch conference on 22/3/86).
3. Bunker et al. Prevalence of retinitis pigmentosa in Maine. Am J Ophthalmol 1984; 97: 357-365.
4. Marmor et al. Retinitis pigmentosa, a symposium on terminology and methods of examination. Ophthalmol 1983; 90: 126-131.
5. Bhattacharia SS, et al. Close genetic linkage between X linked retinitis pigmentosa and a restriction fragment length polymorphism identified by recominant D.N.A. probe L1.28 Nature 1984; 309: 253-255.