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Visual Acuity and 10° Automated Static Perimetry in Eyes with Retinitis Pigmentosa

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Purpose: In a previous study we demonstrated that the progression of the disease retinitis pigmentosa (RP) can be readily monitored by the mean deviation (MD) measured by Humphrey central 10-2 perimetry, which assesses the sensitivity distribution in the macular area in eyes affected by RP. In the present study, we investigated whether the 10° perimetric results could predict the time of declining visual acuity in eyes with RP in a cross-sectional study.

Methods: Humphrey 10-2 perimetry results and visual acuity were studied in the right eyes of 69 patients with typical RP. Patients whose eyes had cataract, glaucoma, cystoid macular edema, or other complications affecting vision were excluded.

Results: Eyes with an MD of −15 dB or greater had almost normal visual acuity. Various degrees of visual acuity loss were observed in eyes with an MD of less than −15 dB. In the 35 eyes with an MD of less than −15 dB, visual acuity correlated well with the corrected pattern standard deviation (CPSD), which is the measure of the degree to which the shape of the measured field departs from the age-corrected normal reference field.

Conclusion: In the absence of complications, many eyes with RP may experience acuity loss after the field constriction reaches an MD of less than −15 dB. The CPSD may be used as an indicator of acuity because eyes showing a lower CPSD tend to have greater loss of acuity among eyes with an equivalent MD value. **Jpn J Ophthalmol 2002;46:581–585** © 2002 Japanese Ophthalmological Society

Key Words: Automated static perimetry, corrected pattern standard deviation, retinitis pigmentosa, visual acuity, visual field.

Introduction

Retinitis pigmentosa (RP) is a hereditary retinal disease characterized by progressive visual field defects. In the typical form of the disease, visual acuity tends to be preserved for many years.[1] Acuity may, however, start to decline at a certain point in the progression of the disease, even without such complications as cataract, glaucoma, or macular edema.[2] Therefore, a major concern for patients with RP is knowing when their acuity will start to decline and how great a reduction of visual acuity to expect. It has been demonstrated that the rate of progression of the disease can be readily assessed in many patients through the annual loss of mean deviation (MD) in the central 10° field.[3] If the visual acuity is closely related to MD or other parameters of static perimetry, we could then more accurately predict the time when the RP patients' central visual acuity will start to decline and advise them more precisely on their visual prognosis.

In the present study we investigated the relationship between perimetric parameters and visual acuity loss in a cross-sectional study.

Materials and Methods

The right eyes of 69 patients (36 men and 33 women, ranging in age from 7 to 88 years with a mean of 50) with the typical form of RP[1] were selected from a database of 194 RP patients registered at the Yama-

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nashi Medical University Hospital. Clinical records were reviewed to determine inheritance patterns and to exclude those patients with other diseases that could cause visual field loss. Excluded were patients with cataract, pseudophakia with posterior capsular opacity, glaucoma, cystoid macular edema, or surface wrinkling of the internal limiting membrane. Patients with retinitis punctata albescence, Leber's congenital amaurosis, choroideremia, and RP syndromes such as Usher syndrome and Bardet-Biedle syndrome, were likewise excluded. The hereditary patterns of the patients included in the study consisted of 11 autosomal dominant, 10 autosomal recessive, 4 X-linked recessive, and 44 simplex cases, including 11 with parental consanguineous marriage.

All eyes were examined with best-corrected visual acuity, by tonometry, slit-lamp biomicroscopy, indirect ophthalmoscopy, and the Humphrey perimetry program central 10-2 with FASTPAC strategy (Carl Zeiss-Humphrey, San Leandro, CA, USA) using stimulus size III and a background of 31.6 asb. The perimetric results used in this study were considered in conjunction with previous examination experience to overcome the learning effect.[4] Visual field data with fixation loss scores of 20% or more and false-positive or false-negative errors of 33% or more were excluded as unreliable in the study following the guidelines presented in the STATPACK user's guide (Allergan-Humphrey, 1986).

Visual acuity was measured using a Japanese standard acuity chart (MT-366; Takagi Seiko, Nakano, Nagano) presented at 5 meters, and converted into a logarithm of the minimum angle of resolution (logMAR). The Humphrey perimetry results were processed with STATPACK for Windows to obtain the MD and the corrected pattern standard deviation (CPSD).[5]

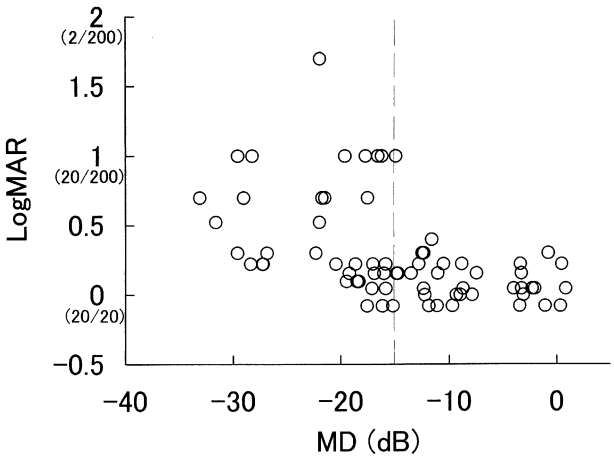

Figure 1. Scattergram displaying the relationship between the mean deviation (MD) of the Humphrey central 10-2 program and the logarithm of the minimal angle of resolution (logMAR) ($r = 0.5$, $P = <.001$). Note that eyes with an MD of −15 dB or higher show good visual acuity.

Results

While logMAR was correlated with MD ($r = 0.5$, $P < .001$), which ranged from −33.1 to 0.9 dB (Figure 1), most eyes with an MD of −15 dB or higher showed a logMAR value of 0.3 or less (corresponding to vision of 20/40 or better). In contrast, eyes with an MD of less than −15 dB showed vision loss of various degrees. Figure 2 shows the relationship between CPSD and MD for eyes assigned to one of three groups according to logMAR. Although the highest value of CPSD depends on MD values, better visual acuity (lower logMAR) is closely related with a higher CPSD value in each level of MD less than −15 dB. To study the relationship between logMAR and CPSD in the relatively advanced stages of the disease, eyes with an MD of −15dB or less were plotted in a scattergram, which revealed that CPSD was negatively correlated with logMAR, meaning that eyes with higher CPSD values tend to have better visual acuity (Figure 3) ($r = 0.45$, $P = .007$).

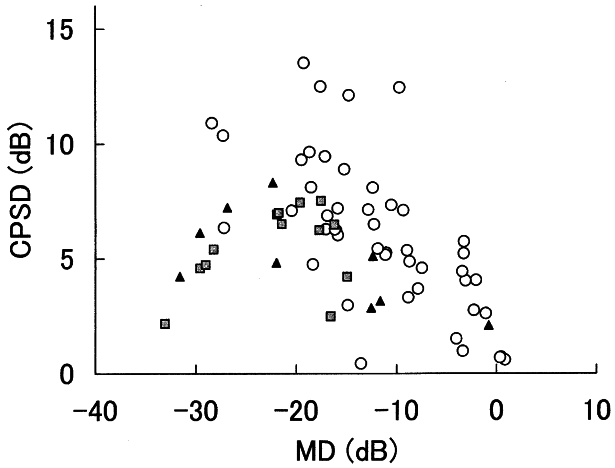

Figure 2. Scattergram displaying the relationship between the mean deviation (MD) and the corrected pattern standard deviation (CPSD) of the Humphrey central 10-2 program. Eyes were assigned to one of three groups according to the level of the logarithm of the minimal angle of resolution (logMAR). ○ logMAR < 0.3 (visual acuity [VA] > 20/40), ▲ 0.6 > logMAR ≥ 0.3 (20/100 < VA ≤ 20/40), ■ logMAR ≥ 0.6 (VA ≤ 20/100).

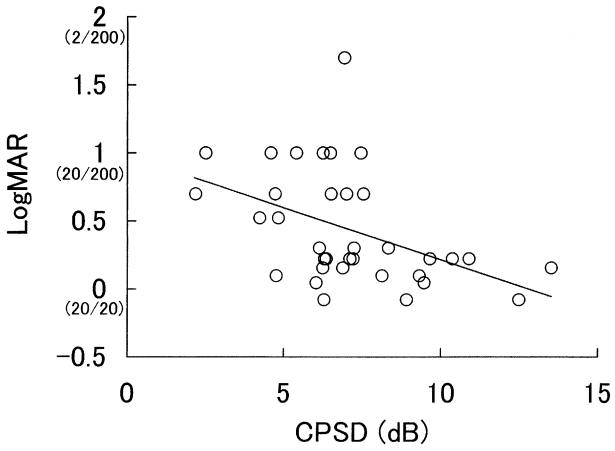

Figure 3. Scattergram displaying the relationship between the corrected pattern standard deviation (CPSD) of the Humphrey central 10-2 program and the logarithm of the minimal angle of resolution (logMAR) in 35 eyes with a mean deviation (MD) of −15 dB or less ($r = 0.45$, $P = .007$).

Discussion

In a previous study,[3] MD measured by Humphrey central 10-2 perimetry decreased linearly in a follow-up period of longer than 3.5 years in eyes with RP. Therefore, MD can be used as a parameter for disease progression, especially in eyes with relatively advanced RP.

The negative correlation between MD and logMAR demonstrated in the present cross-sectional study suggests that visual acuity also decreases with field defect progression in the central 10° visual field. Because eyes with lens opacities, glaucoma, cystoid macular edema, and surface wrinkling of the internal limiting membrane were excluded from the present study, the inherent degenerative changes in the photoreceptors and retinal pigment epithelium in the macula may have been the cause of the acuity loss,[1] which is not apparent until MD decreases to below −15 dB.

Figure 1 shows that the reduction in MD and the visual acuity loss in the disease process are not exactly parallel. For example, an eye with an MD as low as −30 dB still maintained vision of 20/30 (logMAR is 0.2), while another with an MD of −16 dB showed vision as low as 20/200 (logMAR is 1.0).

The total deviation display of the Humphrey central 10-2 results provides a map of sensitivity loss from the age-matched normal value at 68 test points.[5] The histogram of the values in the total deviation display revealed marked differences in pattern between eyes with excellent vision and those with poor vision with equivalent MD values. Figures 4a and 4b show a total deviation plot of the central 10-2 program and a histogram of its individual values for an eye with RP with vision of 20/30. The total deviation plot showed almost normal values higher than −5 dB around the fixation point, and severe reduction of sensitivity less than −30 dB in the periphery of the test field. Such a pattern of perimetric results represents a pencil-shaped "island of visual field." Consequently, the histogram shows a U-shaped pattern with many points located in either the normal area or in that of severe damage. This pattern leads to a value of CPSD as high as 13.5 dB in this case. Figures 5a and 5b show a total deviation plot and a histogram of individual values for an eye with RP with vision of 20/200. The total deviation plot shows almost equally reduced sensitivity throughout the test field, representing a low plateau-shaped "island of visual field." Thus, the histogram shows an inverse U-shaped pattern with most points being concentrated in the area of moderately reduced sensitivity. This pattern leads to a CPSD value as low as 2.7 dB in this case.

The present results showing inverse correlation between CPSD and logMAR (Figure 3) could be interpreted as follows: eyes with a pencil-shaped field island in the central 10° field, such as that shown in Figure 4a, tend to show good visual acuity until an advanced stage of the disease when MD values become very low, while eyes with a low plateau-shaped island of visual field, such as that shown in Figure 5a, tend to lose acuity at an earlier stage in which MD shows a moderate reduction of around −15 dB.

We do not know at present what pathological changes may result in these two patterns. Cones in the center of the macula might be more resistant to the degenerative changes that occur in the progression of the disease in eyes with a pencil-shaped field island. The vision loss may be due to degenerative changes in the foveal photoreceptors and retinal pigment epithelium cells, which would result in window defects in the angiogram of the macular area,[6,7] although we did not perform fluorescein fundus angiography on all subjects.

Recently, many different mutations in the genes expressed in the photoreceptors have been found to cause RP. A study of the genotype-phenotype relationship has revealed that the particular genetic mutations could explain differences in the severity of clinical dysfunction.[8–10] Because we have not studied the causal mutations in the gene we are unable to determine whether the two different patterns in the cen-

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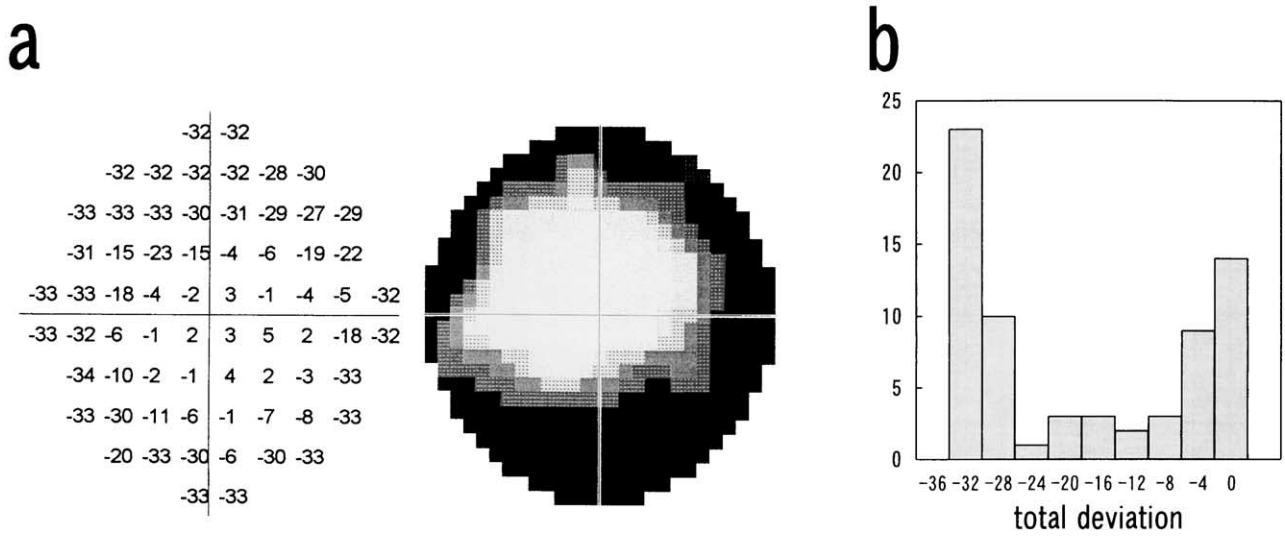

Figure 4. (**a**) Total deviation and grayscale display of an eye with retinitis pigmentosa with 20/30 vision. The values of the points around the fixation point are almost normal, while those situated in the periphery of the 10° field are severely reduced. (**b**) Histogram of the sensitivity loss from normal values in the total deviation display for the eye shown in Figure 4a. The corrected pattern standard deviation is 13.5.

tral 10° field might result from differences in genetic disorder. This problem remains to be investigated.

In summary, we have demonstrated a close relationship between visual acuity and the parameters of central 10° automated perimetry in eyes with uncomplicated RP. Decreased visual acuity is usually found in eyes with an MD of less than −15 dB, in which higher CPSD is related to better visual acuity.

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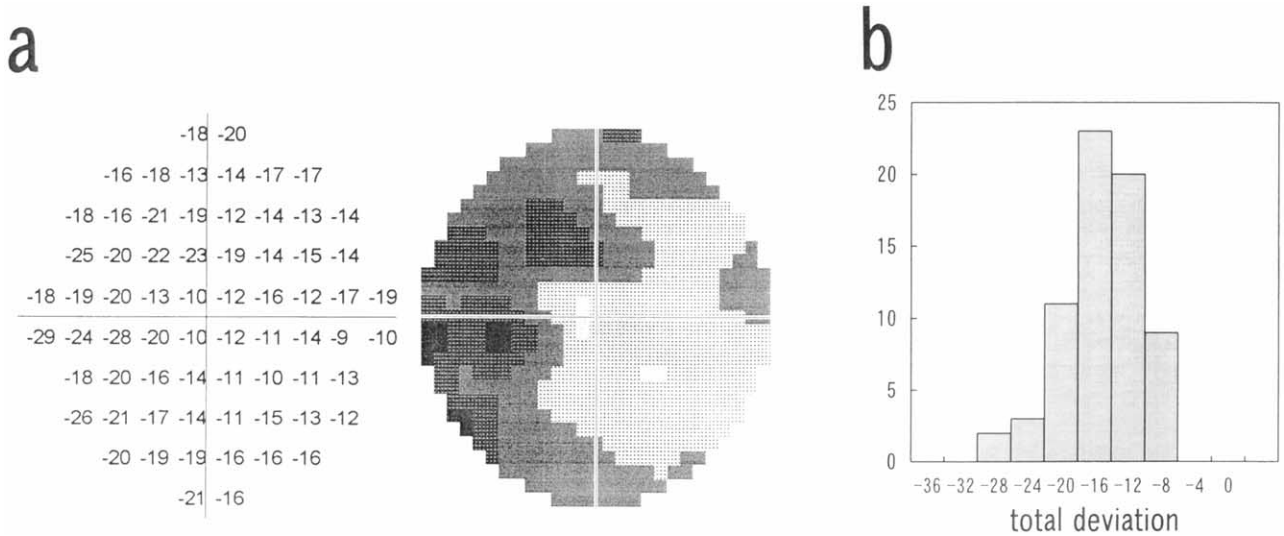

Figure 5. (**a**) Total deviation and grayscale display of an eye with retinitis pigmentosa with 20/200 vision. The values of the points around the fixation point are moderately reduced, showing little difference from those in the periphery in the 10° field. (**b**) Histogram of the sensitivity loss from normal values in the total deviation display for the eye shown in Figure 5a. The corrected pattern standard deviation is 2.7.

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