Figure legends

Figure 1. The distribution of age in highly myopic patients with PDPM (hatched bar) and without PDPM (black bar).

Figure 2. The distribution of refractive errors in highly myopic patients with PDPM (hatched bar) and without PDPM (black bar).

Figure 3. The distribution of axial length measurements in highly myopic patients with PDPM (hatched bar) and without PDPM (black bar).

Figure 4. Case 1. A 60-year-old man. **A and B:** The right fundus shows a yellow inferonasal peripapillary elevated lesion at the inferior edge of the optic disc and conus, clearly distinct from the myopic conus. There is a deep excavation in the inferior part of the myopic conus, and inferotemporal retinal vein was markedly bent at the border edge between the myopic conus and PDPM (arrowhead). **C:** Fluorescein fundus angiogram in the early phase reveals hypofluorescence at the PDPM area (arrow). **D:** Fluorescein fundus angiogram in the late phase shows hyperfluorescence at the PDPM area (arrow). **E:** The OCT scan across the PDPM (arrow in Figure 4B) shows a nonreflective area that appears to be beneath the RPE and retina corresponding to the PDPM area (white arrow). The left side of the scan is the optic disc. **F:** Goldmann kinetic perimetry reveals arcuate scotoma, nasal step, and mild temporal wedge, which are not explained by myopic fundus changes.

Figure 5. Case 2. A 68-year-old man. **A:** The left fundus shows a yellow peripapillary elevated lesion almost all around the optic disc, clearly distinct from the myopic conus (arrowhead). There is a deep excavation in the inferior part of the myopic conus, and the inferotemporal retinal vein was strongly bent at the border of myopic conus and PDPM (arrow). Choroidal neovascular membrane with pigmentation is observed in the macula. **B:** The OCT scan across PDPM (Figure 5A, white arrow) shows a nonreflective area beneath both the RPE and retina corresponding to the PDPM area. The left side of the figure shows the optic disc. **C and D:** Fluorescein fundus angiogram of the left fundus. A fluorescein angiogram reveals early hypofluorescence (Figure 5C) and the late staining (Figure 5D) at the PDPM area. **E and F:** ICG angiograms of the left fundus. ICG angiogram demonstrates that the inferotemporal retinal vein enters into the PDPM space (arrowhead) and eventually flows back into the center of the optic disc (Figure 5E, arrowhead). ICG angiogram demonstrates mild hypofluorescence at the PDPM area in the late phase (Figure 5F, arrowhead).

Figure 6. Case 3. A 48-year-old man. **A:** The right fundus shows a small, yellow inferonasal peripapillary elevated lesion at the inferior edge of the optic disc (arrowhead). **B:** The OCT across the PDPM (Figure 6A, arrow) shows a nonreflective area beneath both the RPE and retina corresponding to the area of PDPM. An OCT scan through the PDPM reveals what appears to be a full-thickness defect in the retina-RPE layers (white arrow). **C:** Goldmann kinetic perimetry

reveals a nasal step and arcuate scotomas, which are not explained by myopic fundus changes. **D:** Humphrey C30-2 program demonstrates arcuate scotomas and nasal step.

Figure 7. Case 4. A 53-year-old man. **A:** The right fundus shows a yellow peripapillary elevated lesion mainly situated superotemporal to the optic disc in addition to a small lesion inferior to the optic disc. There is a deep excavation of the myopic conus, and the inferior retinal veins seem to disappear at the edge of inferior myopic conus (white arrows).**B:** Goldmann kinetic perimetry reveals a nasal step and arcuate scotomas that are not explained by myopic fundus change. **C and D:** ICG angiograms of the right fundus. ICG angiograms demonstrate mild hypofluorescence at the area of PDPM from the early phase (7C) through the late phase (7D). **E:** The OCT across the PDPM (Figure 7A, arrow) shows a nonreflective area beneath both the RPE and retina corresponding to the area of PDPM. An OCT scan through the PDPM reveals what appears to be a full-thickness defect in the retina-RPE layers (arrow).

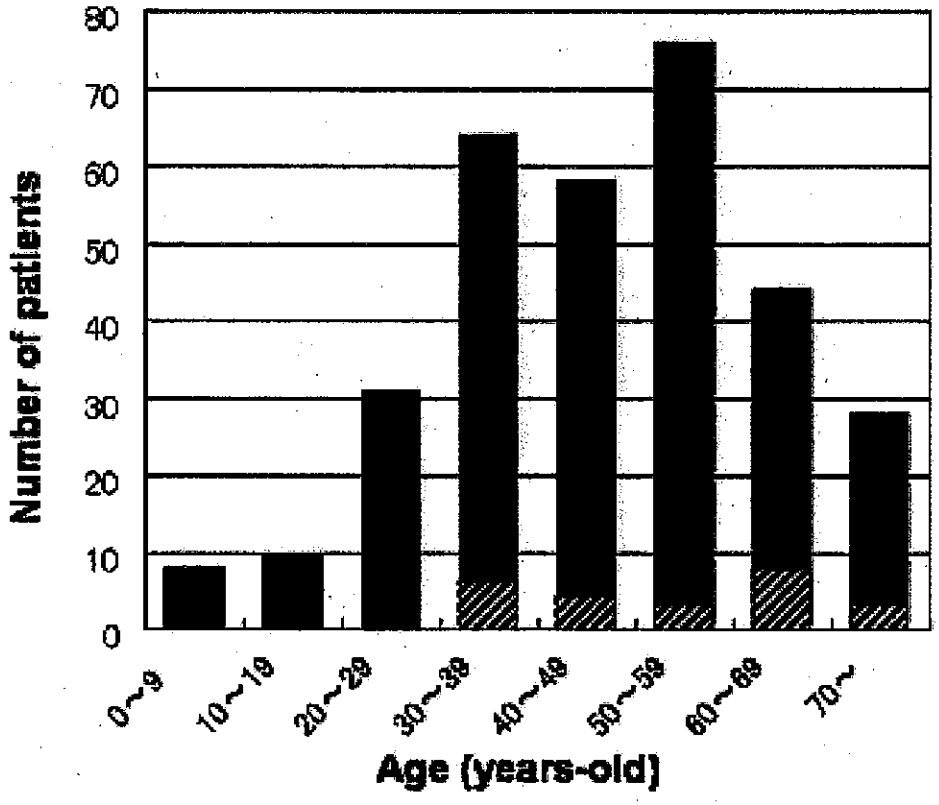
80
70
60
50
40
30
20
10
0
Number of patients
0~9
10~19
20~29
30~39
40~49
50~59
60~69
70~
Age (years-old)

Figure 1

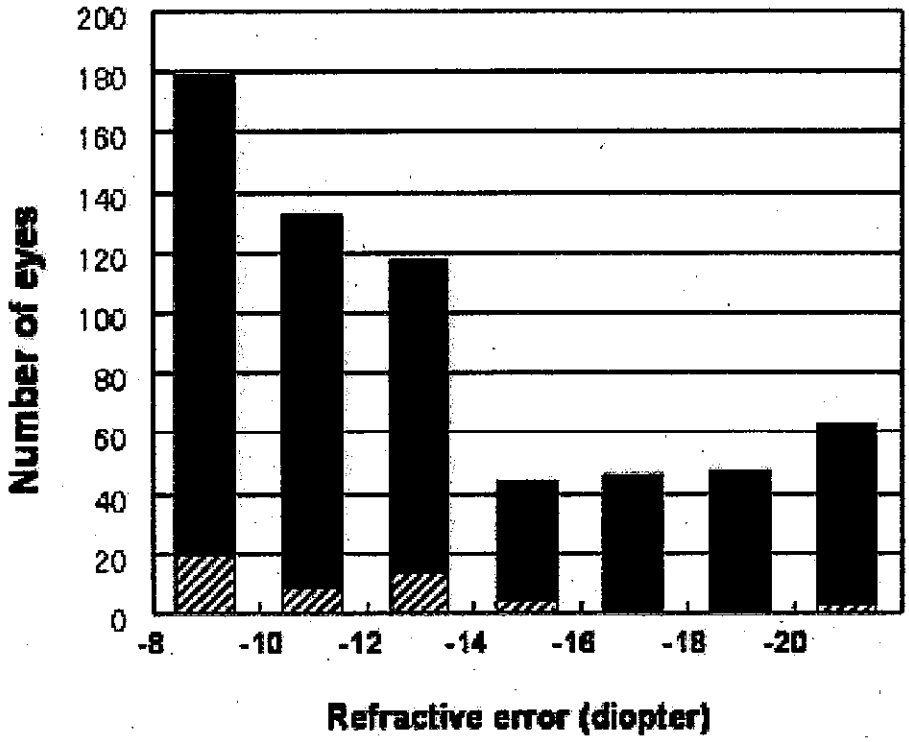
200
180
160
140
120
100
80
60
40
20
0
Number of eyes
-8
-10
-12
-14
-16
-18
-20
Refractive error (diopter)

Figure 2

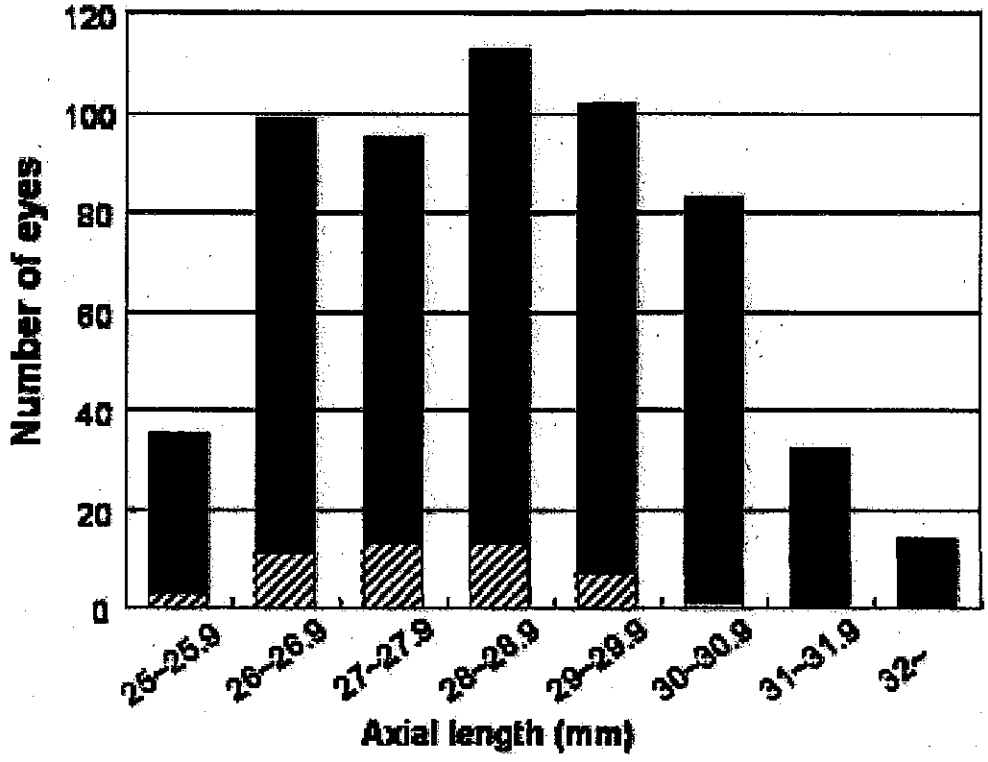
120
100
80
60
40
20
0
Number of eyes
25~25.9
26~26.9
27~27.9
28~28.9
29~29.9
30~30.9
31~31.9
32~
Axial length (mm)

Figure 3

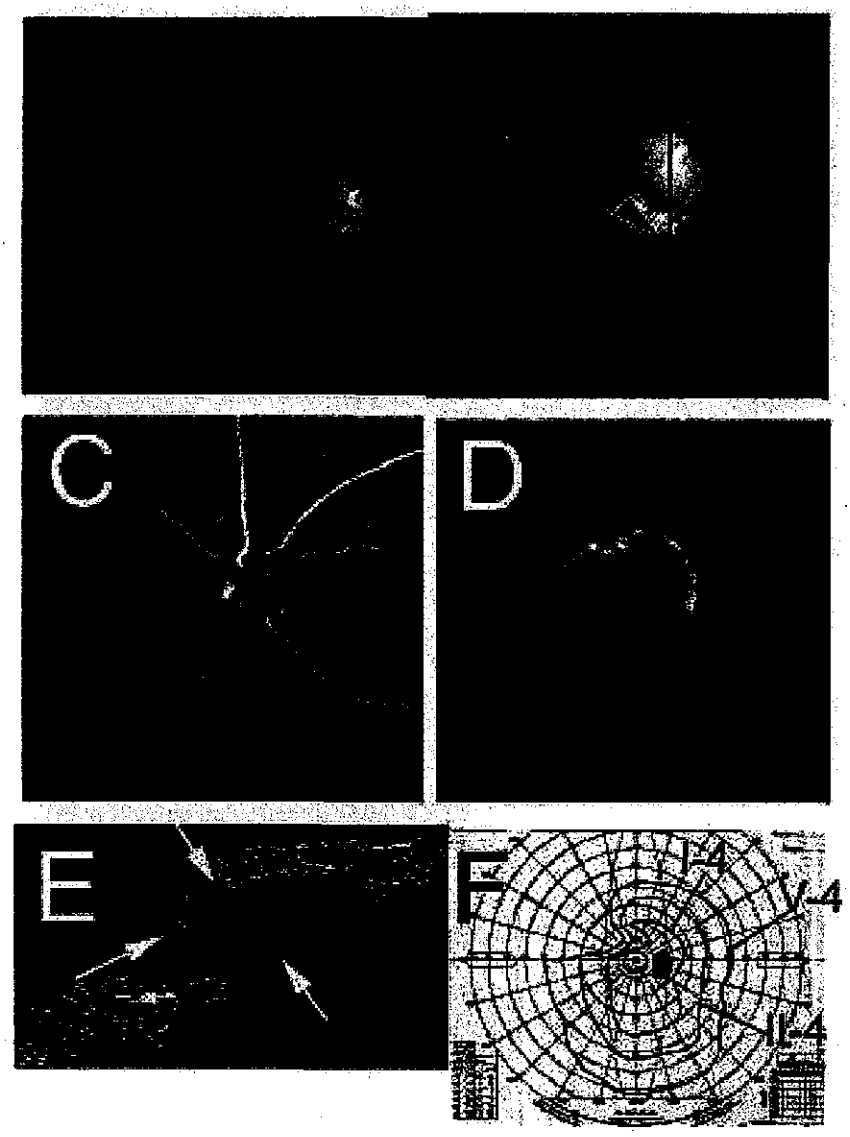

Figure 4

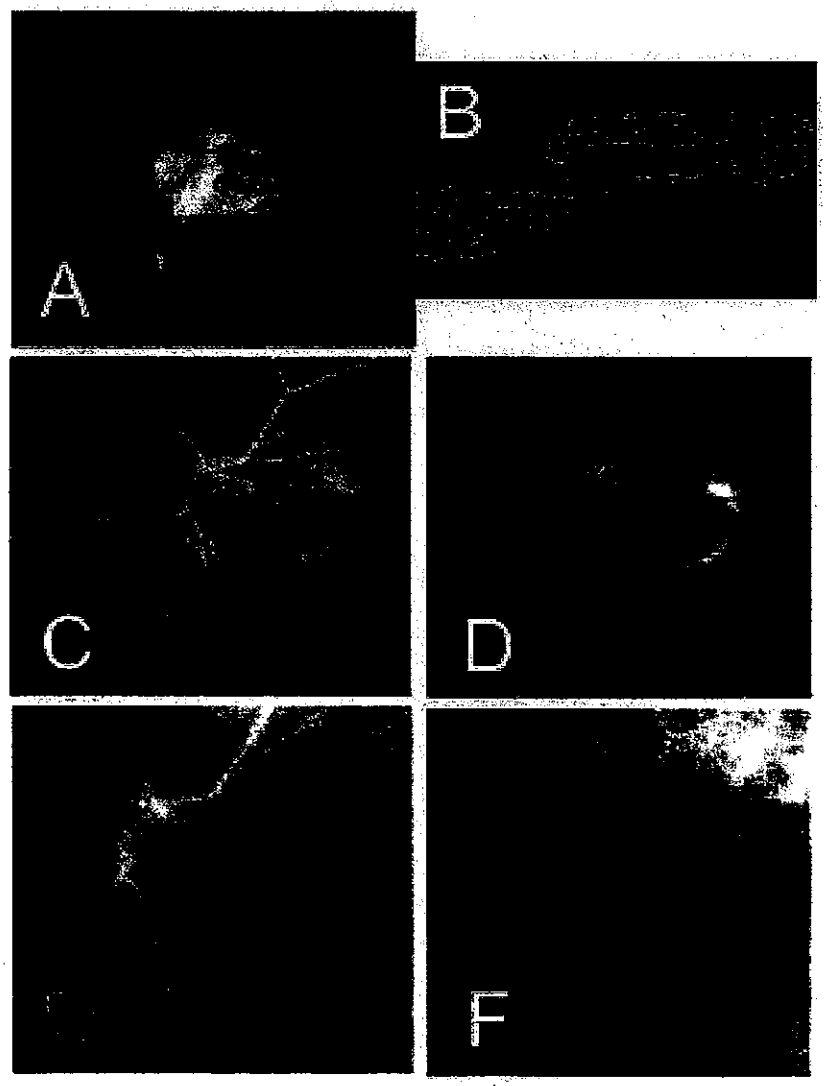

Figure 5

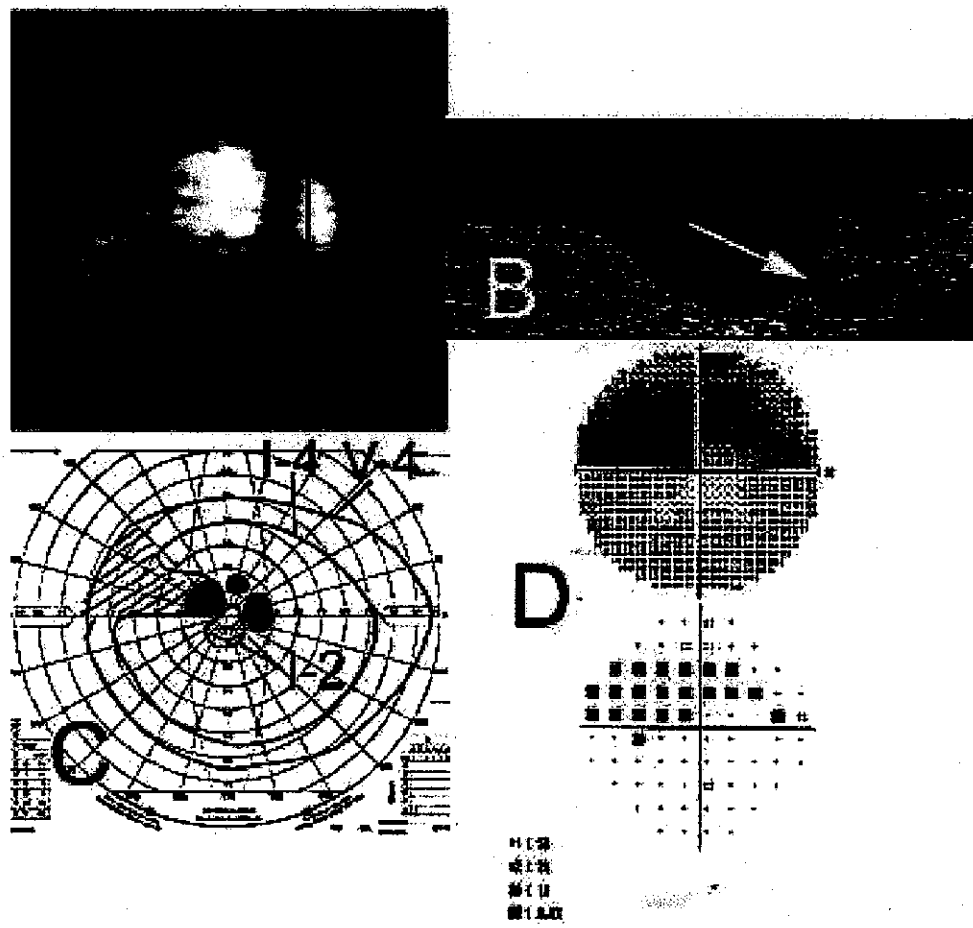

Figure 6

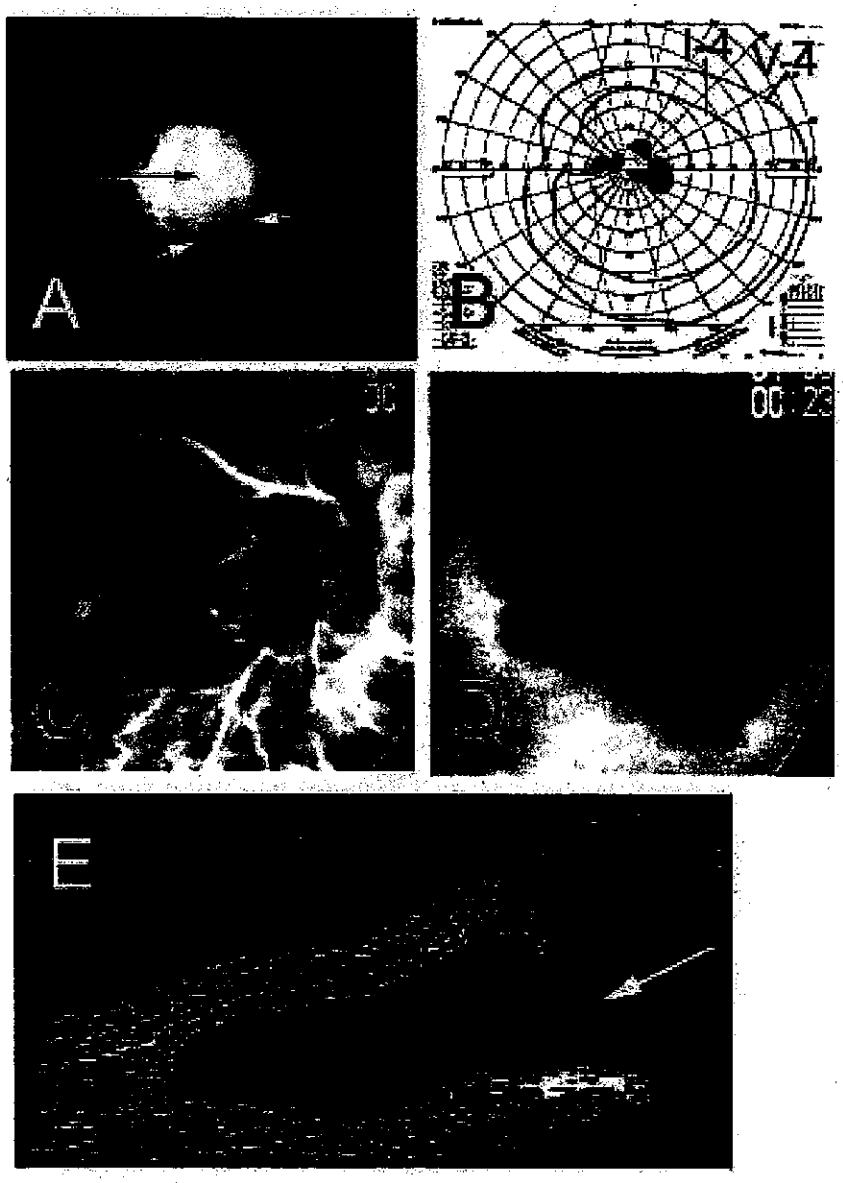

Figure 7

Table 1. Patient and Study Eye Characteristics

Gender, no. persons (eyes)	23 (31)
Men	13 (17)
Women	10 (14)
Age (yrs), mean (SD)	57.7 (14.3)
logMAR, mean (SD)	0.59 (0.34)
Refractive error (D), mean (SD)	-11.4 (3.53)
Axial length (mm), mean (SD)	27.7 (1.42)
Tilted discs	29/31 (93.5%)
Peripapillary crescent	31/31 (100%)
Posterior staphyloma	19/31 (61.3%)

Fundus characteristics of high myopia in children

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Abstract

Aim: To evaluate the fundus characteristics of highly myopic eyes in children.

Methods: Medical records of 80 eyes of 46 children (1 to 8 years; mean age 6.8 years) with high myopia (4 D or more for children younger than 5 years, 6 D or more for children aged between 6 to 8 years) seen consecutively during a 10 - year period were reviewed. The age was limited to less than 8 years.

Results: Fundus examination revealed posterior staphyloma in only one eye (1.3%) and mild chorioretinal atrophy around the optic disc in 13 eyes (16.3%).There were no patients with choroidal neovascularization or geographic atrophy in the posterior fundus. Myopic peripapillary crescent was observed in 26 eyes (33.8%), but the area of the crescent was relatively small (mean; 0.5 disc area).

Conclusion: The results of the present study indicated that myopic fundus changes are uncommon and mild in children. This suggests that aging, in addition to mechanical stretching of the eyeball, might be important for the development of myopic fundus changes.

Introduction

High myopia is a major cause of legal blindness in many developed countries.[1,2] It affects 27 to 33% of all myopic eyes, corresponding to a prevalence of 0.2 to 0.4% in the general population of the United States.[3] High myopia is especially common in Asia and the Middle East.[2] Although the frequency of affected patients in the general Japanese population is not clear, it is estimated at approximately 1%.[4] Since myopic eyes with a refractive error greater than -8.0 diopters are frequently associated with visual dysfunction, high myopia is defined as an eye with a refractive error greater than -8.0 diopters, and this definition has been used as a definition criteria for high myopia in Japan.[4]

High myopia is associated with progressive and excessive elongation of the eyeball, which results in various funduscopic changes in the posterior fundus.[5,6] These changes include areas of atrophy of the retinal pigment epithelium and choroid, lacquer cracks in Bruch's membrane, subretinal hemorrhage, and choroidal neovascularization. Also, a large peripapillary crescent is a hallmark of highly myopic eyes.[7]

The clinical findings suggest that the prevalence of these myopic fundus changes increases gradually as patients age. Curtin and Karlin[7] demonstrated that

the incidence of chorioretinal atrophy in highly myopic eyes increases in patients 40 years and older, which suggests that myopic fundus changes might be rare in children. High myopia in children is rare. Lin et al[8] reported that the prevalence of high myopia (>-6.0D) was 0.2% at 7 years-old among 10889 children in Taiwan. Also, most other studies of children with high myopia have focused only on treatment and visual outcome. To our knowledge, there are no detailed reports evaluating the fundus characteristics in highly myopic eyes of children. In the present study, we examined the characteristics of myopic fundus changes in the posterior pole (especially myopic crescent, posterior staphyloma, and chorioretinal atrophy) in highly myopic eyes in children.

Patients and Methods

Medical records of 80 eyes of 46 children (1 to 8 years; mean age 5.4 years) with high myopia were reviewed. The study sample consisted of all children (younger than 8 years at the initial visit) seen consecutively over a 10 - year period among patients who visited high myopia clinic in Tokyo medical and dental university from 1980 to 1990. High myopia was defined as eyes with axial length exceeding three times the standard deviation from a normal distribution of the emmetropic axis.[9] When the refractive degree of the eye is calculated from the long axial length outside its

normal distribution, high myopia is determined when the measurement of refraction exceeds -4.0 D for children younger than 5 years and -6.0 D for children between 6 and 8 years.[9]

All patients had complete eye examinations. Informed consent was obtained from all the patients. Refractive errors (degrees) were determined 40 minutes after instilling one or two drops of 1% cyclopentolate. Patients who were younger than 2 years were examined by streak retinoscope or hand refractometer (Retinomax, Nikon, Tokyo, Japan) and those who were older than 2 years were examined using an autorefractometer (AR-600A, Nidek, Aichi, Japan). Spherical equivalents of refractive status were used. Total axial length (AL) was measured by A-scan ultrasonography (Ultrascan, Alcon, Fort Worth, TX). Fundus examination was performed using stereoscopic indirect ophthalmoscopic evaluation. The chorioretinal atrophy grade was assigned according to Steidl et al.[6] Grade 0 was given to eyes without evidence of atrophic change. Grade 1 eyes had attenuated choroidal vessels, limited lacquer crack formation, and retinal pigment epithelial mottling, or a combination of these. Grade 2 eyes had a total area of geographic atrophy less than or equal to 2 disc areas. Grade 3 eyes had a total area of geographic atrophy greater than 2 disc areas but less than or equal to 4 disc areas. Grade 4 eyes had a total area of geographic

atrophy greater than 4 disc areas. Area measurements were performed using color photographs. The measurement of the area of the myopic crescent was described previously.[10] Briefly, fundus photographs were scanned using an image scanner (Scanjet CX/T, Hewlett Packard; Palo Alto, CA), and exported to the public domain NIH Image program (version 1.62 ; developed at the U.S. National Institutes of Health and available on the Internet at http://rsb.info.nih.gov/nih-image/). The area of the peripapillary crescent was defined as the ratio of the total number of pixels of the crescent area divided by the number of pixels of the optic disc area (DA).

Visual acuities were measured using a Dot visual acuity cards[11] or a standardized visual acuity chart using Landolt Cs depending upon the patient's age and ability level. To provide a common measure and linear scale, decimal notations were transformed into the logarithm of the minimum angle of resolution in minutes of arc (logMAR).

Results

Eighty eyes of 46 patients (34 males, 12 females) ranging from 1 year to 8 years of age (mean age 5.4 ± 2.1 years) satisfied the selection criteria. Figure 1 shows the age distribution among the patients. Figure 2 shows the refractive errors in diopters for the 80 eyes. The mean refractive error was -8.4 ± 3.8 D. Figure 3 shows

the AL values in millimeters for the 60 eyes in which AL measurement was obtained. The mean axial length was 25.7 ± 9.8 mm. Thirty eyes had AL longer than 25.5 mm. Visual acuity measurements were performed in 45 patients. The mean logMAR was 0.37 ± 0.41 (range -0.18 to 1.30).

Three patients had ocular or systemic abnormalities. One patient had encephalitis, one had lid hemangioma, and another had widespread myelinated nerve fibers in the posterior fundus. Twenty-three patients (50%) had a family history of high myopia (within 2 pedigrees).

Fundus examination revealed that only 13 eyes (16.3%) had Grade 1 chorioretinal atrophy. No patients had chorioretinal atrophy more severe than Grade 2. Also, no patients had choroidal neovascularization. Posterior staphyloma was detected in only one eye (1.3%). Myopic peripapillary crescent was observed in 26 eyes (33.8%; mean area 0.46 ± 0.27 DA; range: 0.17 to 1.37 DA). When we limit the patients to those with an axial length longer than 25.5 mm (30 eyes), posterior staphyloma was detected in 1 eye (3.3%), chorioretinal degeneration in 7 eyes (23.3%), and myopic crescent in 14 eyes (46.7%).

Case Reports

Case 1. A 6-year-old girl was referred to our hospital because of high myopia in both

eyes in June, 1995. At the initial examination, the patient's best-corrected visual acuity was 0.4 in the right eye and 0.2 in the left eye. The refractive error was -5.75 D in the right eye and -7.75 D in the left eye, and the axial length measurements were 24.4 mm in the right eye and 25.2 mm in the left eye. There was yellowish chorioretinal atrophy around the optic discs of the fundus in both eyes (Fig. 4).

Case 2. A 7-year-old girl was referred to our hospital because of high myopia in the right eye in May, 1994. At the initial examination, the patient's best-corrected visual acuity was 0.1 in the right eye and 1.5 in the left eye. The refractive error was -16.5 D in the right eye and +0.5 D in the left eye, and the axial length measurements were 25.5 mm in the right eye and 22.3 mm in the left eye. There was a posterior staphyloma nasal to the optic disc in the right fundus (Fig. 5).

Discussion

In the present study, we investigated clinical characteristics of children with high myopia. Most of our patients did not have any systemic or ocular complications that might be associated with the development of myopia. Only three children had complications; encephalitis, lid hemangioma, or widespread myelinated nerve fibers. Central nervous system abnormalities,[12] obstruction within the visual axis (lid ptosis, cataract),[12] and myelinated nerve fibers[13] are all causes of axial myopia.

Pathologic myopia can be accompanied by a number of visually disabling complications.[5] Hallmarks of the process include lacquer cracks in Bruch's membrane, choroidal neovascular invasion of the macula, and areas of choriocapillary and pigment epithelial atrophy within the posterior staphyloma. Several previous studies reported the prevalence of myopic fundus changes in highly myopic patients (Table 1). These studies include patients from all ages, however, and the prevalence of myopic fundus changes among only children is not clear. Gozum et al.[14] examined 212 eyes of 109 patients (age; 9 to 70 years, mean 31 years) with high myopia and reported that posterior staphyloma was detected in 23.6%, Fuchs' spot in 6.6%, and peripapillary crescents in 66.5% of the eyes. Curtin[7] reported a 75% incidence of myopic crescent in eyes with an axial length of 24.5 to 25.4 mm and increased to 100% above 28.5 mm in 1437 eyes from all age groups. Tekiele et al.[15] examined 56 myopic eyes with a refractive error greater than 6.00 D or greater. An optic nerve head crescent occurred in 17 eyes (30.4%). Grossniklaus and Green[16] examined 308 postmortem eyes with pathologic myopia obtained from 202 patients histopathologically. The patients' age ranged from 3.5 to 93 years. The results indicated that myopic configuration of the optic nerve head was observed in 37.7%, posterior staphyloma in 35.4%, myopic degeneration of the retina in 11.4%, and