CrossMark

American Academy of Ophthalmology
The Eye M.D. Association

Comparison of Clinical Features in Highly Myopic Eyes with and without a Dome-Shaped Macula

I-Chia Liang, MD,[1,2] Noriaki Shimada, MD, PhD,[1] Yuichiro Tanaka, MD,[1] Natsuko Nagaoka, MD,[1] Muka Moriyama, MD, PhD,[1] Takeshi Yoshida, MD, PhD,[1] Kyoko Ohno-Matsui, MD, PhD[1]

Purpose: To compare the clinical features of highly myopic eyes with a dome-shaped macula (DSM) with those without a DSM and to identify the funduscopic clues to suspect the presence of DSM.

Design: Retrospective case series.

Patients: A total of 586 patients (1118 eyes) with high myopia (refractive error <-8 diopters [D] or axial length ≥ 26.5 mm) who had optical coherence tomography (OCT) examinations through the central fovea at our High Myopia Clinic between February 2012 and November 2013.

Methods: Vertical and horizontal OCT scans across the central fovea were retrospectively analyzed. A DSM was defined by the presence of an inward bulge of >50 μm in the vertical OCT image. Fundus photographs also were analyzed to identify the funduscopic clues to suspect the presence of DSM.

Main Outcome Measures: The rate of DSM in 1118 highly myopic eyes. The rate of DSM in highly myopic eyes with macular complications. Funduscopic features to suggest the presence of DSM.

Results: Among the 1118 eyes, 225 (20.1%) had a DSM. A DSM was present in both vertical and horizontal OCT sections in 20% of eyes, along only the vertical section in 77% of eyes, and in only the horizontal section in 2% of eyes. The results of multiple regression analyses showed that serous retinal detachment and foveal and extrafoveal retinoschisis were significantly associated with the presence of DSM and that choroidal neovascularization was not. Ophthalmoscopically, 91.4% of the eyes with the appearance of a horizontal ridge connecting the optic disc and the fovea had a DSM.

Conclusions: A DSM is found in as many as 20% of highly myopic individuals. Horizontal ridges connecting the optic disc and the fovea might be an important clue to suspect the presence of a DSM on the basis of fundus photographs. *Ophthalmology 2015;122:1591-1600 © 2015 by the American Academy of Ophthalmology*

Supplemental material is available at www.aaojournal.org.

The dome-shaped macula (DSM) was first described by Gaucher et al[1] as a convex protrusion of the macula in highly myopic eyes detected in optical coherence tomography (OCT) images. Imamura et al[2] used enhanced-depth imaging OCT and showed that the DSM resulted from a relative localized thickening of the sclera under the macula in highly myopic eyes. A DSM has been reported to be associated with various kinds of macular complications, such as a serous retinal detachment (RD),[1,3–7] choroidal neovascularization (CNV),[1,3,4,6,7] and retinoschisis (RS).[4]

However, there are still some uncertainties regarding DSM. First, the rate of DSM among highly myopic patients is not known. Thus, whether DSM is a frequent feature among highly myopic eyes is uncertain. Second, it is not clear whether various macular complications reported to occur in eyes with DSM are unique to DSM or are common complications of highly myopic eyes regardless of the presence of DSM.

A PubMed search using the keyword "dome-shaped macula" on February 10, 2015, extracted 16 articles, with those on tilted disc syndrome excluded.[1–16] However, most of them were case series or case reports examining only cases with DSM. Among these studies, only 2 examined highly myopic eyes with and without DSM. Imamura et al[2] compared the scleral thickness of 23 highly myopic eyes with DSM with that of 25 highly myopic eyes without DSM. Chebil et al[15] reported that a DSM was found in 24 of 200 highly myopic eyes (12%), and they compared the choroidal thickness in eyes with and without DSM. However, the number of the patients with DSM in these studies was still not high.

The High Myopia Clinic at Tokyo Medical and Dental University has been functioning for more than 40 years, and more than 3600 patients have been registered. The purpose of this study was to analyze the frequency and features of DSM in a large series of highly myopic patients. The rate of various macular complications that had been reported to occur in eyes with DSM was compared with those without DSM. In addition, new fundus findings suggesting the presence of DSM were analyzed. Although it was difficult to suspect the presence of DSM from fundus photographs, we

http://dx.doi.org/10.1016/j.ophtha.2015.04.012
ISSN 0161-6420/15

believe that it would be useful to determine the clues in fundus photographs to suspect DSM because OCT is not always available in all clinics.

Methods

The institutional review board and ethics committee of the Tokyo Medical and Dental University approved this retrospective study. The techniques used to collect the data conformed to the tenets of the Declaration of Helsinki.

Patients

The medical records of consecutive patients with high myopia at the High Myopia Clinic of Tokyo Medical and Dental University were reviewed, and the cases with vertical and horizontal OCT images through the central fovea were studied. High myopia was defined as a refractive error of ≥ -8.0 diopters, an axial length of ≥ 26.5 mm, or both. There were no specific visual or clinical criteria for being registered in the High Myopia Clinic apart from having high myopia. The eyes that had undergone vitreoretinal surgery were excluded, and those whose OCT images were not clear because of the media opacities were also excluded. In addition, eyes diagnosed with an inferior staphyloma due to congenital tilted-disc syndrome on the basis of stereoscopic fundus examination were excluded.

Optical Coherence Tomography Examinations

The OCT images were obtained with a swept-source OCT (DRI-OCT; Topcon Corp, Tokyo, Japan). For the DRI-OCT images, averaged horizontal and vertical raster scans of 12 mm were recorded. The final image was the average of 1024 scans. For the patients who were not examined by swept-source OCT, the images obtained with the RS3000 (Nidek Co. Ltd, Aichi, Japan) or the Cirrus HD-OCT (Carl Zeiss Meditec, Inc, Dublin, CA) were used. For the RS3000 instrument, 6 radial scans 9 mm in length and centered on the fovea at 0°, 30°, 60°, 90°, 120°, and 150° were obtained. For the Cirrus OCT, vertical and horizontal line scans 6 mm in length and centered on the fovea were obtained. The technique of the enhanced-depth imaging OCT[17] was used with the RS3000 and Cirrus OCT instruments. A single experienced examiner performed the OCT examination while masked to the clinical diagnosis of the subjects.

A DSM was defined as the presence of an inward bulge of the macular retinal pigment epithelium (RPE) of >50 μm in the vertical or horizontal section of the OCT image, according to Ellabban et al[4] and Ohsugi et al.[7] The dome height was measured as the distance between the peak of the bulge to a line tangential to the RPE. The choroidal thickness was defined as the distance from

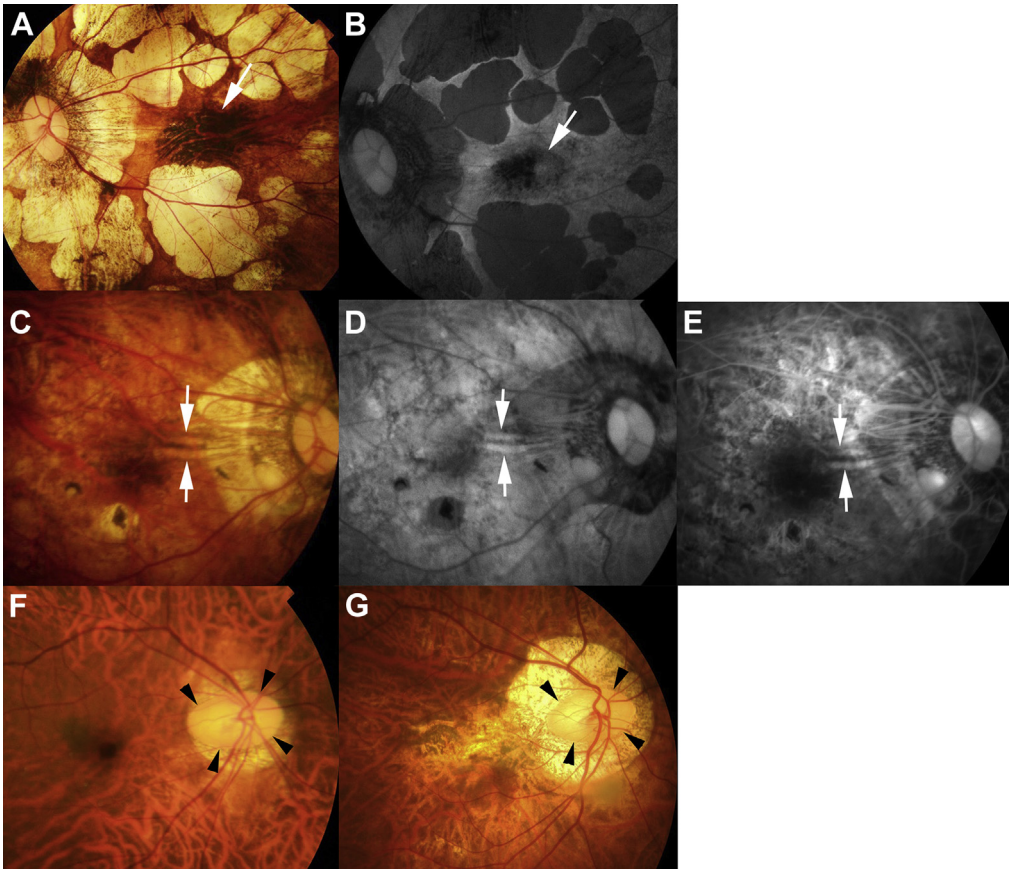

Figure 1. Three fundus lesions as clues that a dome-shaped macula (DSM) might be present. **A, B,** Macular pigmentation (*arrow*) is seen funduscopically as increased pigmentation **(A)** and as hypoautofluorescence **(B)**. **C–E,** Horizontal ridge–like lesion connecting the optic disc and the fovea (between *arrows*). The horizontal ridge is observed funduscopically as a yellowish-white linear lesion **(C),** as hyperautofluorescent **(D)**, and as hyperfluorescent **(E)**. In stereoscopic observations, the horizontal ridge seems to slightly protrude toward the vitreous. **F, G,** Horizontally oval optic disc (*arrowheads*).

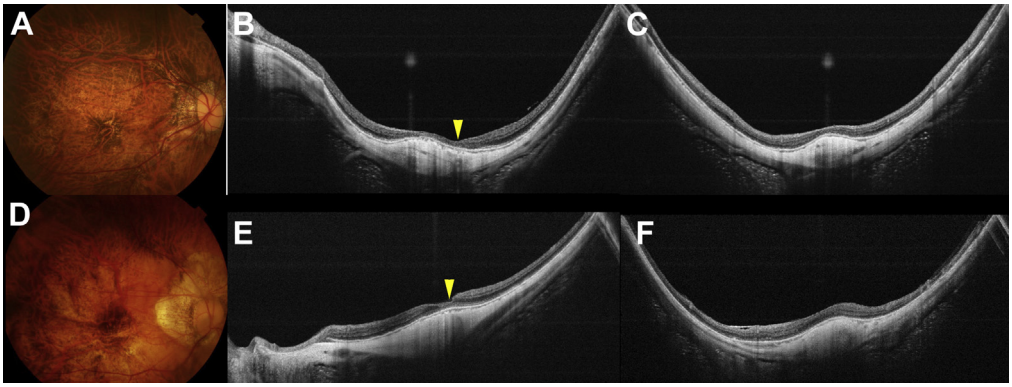

Figure 2. A dome-shaped macula (DSM) seen in both vertical and horizontal optical coherence tomography (OCT) sections through the fovea (bidirectional DSM). **A,** Right fundus of a 62-year-old woman with an axial length of 33.2 mm. **B, C,** Optical coherence tomography images across the central fovea. The sclera is thickened in the macula and shows an inward bulge in the horizontal section **(B)** and vertical section **(C)**. The fovea (*arrowhead* in **B**) is located temporal to the top of the dome in the horizontal section. **D,** Right fundus of an 80-year-old woman with an axial length of 29.7 mm. **E, F,** Optical coherence tomography image across the central fovea. The sclera is thickened in the macula and shows an inward bulge in both the horizontal **(E)** and vertical **(F)** sections. The fovea (*arrowhead* in **E**) is located slightly temporal to the top of the dome in the horizontal section.

the RPE line to the hyperreflective line behind the large vessel layers of the choroid, which was assumed to be the choroid–sclera interface. The subfoveal choroidal and scleral thicknesses were measured by a single masked author (I.-C.L.).

Other Examinations

All of the participants had a comprehensive ocular examination including refractive error measurements with an autorefractometer (ARK-730; Nidek Co. Ltd) without cycloplegia, and axial length measurements using the IOLMaster (Carl Zeiss, Tubingen, Germany). The axial length measurements were routinely performed for all of the patients. Best-corrected visual acuity was determined with a Landolt C chart and was converted to logarithm of minimal angle of resolution units for statistical analyses. Fundus photographs and fundus autofluorescence (FAF) images were obtained using a TRC50LX (Topcon Corp.) or an Optos 200Tx scanning laser ophthalmoscope (Optos PLC, Dunfermline, Scotland, UK). In some patients, infrared images of the retina were obtained by the Optos 200Tx.

Complications of Dome-Shaped Macula

We also analyzed the incidence of various macular complications, such as CNVs, serous RDs, lamellar macular holes, full-thickness macular holes, foveal RS, and extrafoveal RS. The eyes with CNVs were not included in the number of eyes with a serous RD. The diagnosis of CNV was made by a combination of OCT and fluorescein angiography in all cases. Myopic CNVs were classified into 3 phases: active phase, scar phase, and atrophic phase.[18–20] All of the 3 phases were regarded as myopic CNV.

To determine the clues in fundus photographs that would lead clinicians to suspect the presence of DSM, we focused on 3 lesions: macular pigmentation, the appearance of a horizontal ridge that ran from the optic disc to the fovea, and a horizontally oval optic disc (Fig 1). Macular pigmentation is observed as increased pigmentation funduscopically and as hypoautofluorescence in FAF images (Fig 1A and B). Eyes with a history of CNV were excluded when examining whether macular pigmentation was present because myopic CNVs frequently form pigmented scars known as Fuchs' spots. To differentiate macular pigmentation from Fuchs' spots, OCT images were used to detect the presence of subretinal scar tissue due to Fuchs' spots. The appearance of a horizontal ridge connecting the optic disc and the fovea is funduscopically seen as yellowish-white linear lesions (Fig 1C). In stereoscopic observations, the horizontal ridge seems to protrude slightly toward the vitreous, and the ridge was continuous to the area of myopic conus. It often consisted of 2 linear protrusions that ran in parallel, and the ridges did not extend beyond the fovea, unlike the upper border of the inferior staphyloma. The horizontal ridge was hyperautofluorescent in the FAF images (Fig 1D) and hyperfluorescent in the fluorescein angiography images (Fig 1E). The horizontal ridge was also hyperreflectant in infrared images, suggesting an increased reflex from the protruded sclera.

The shape of the optic disc was analyzed with a slight modification of our earlier study.[21] Briefly, when the ratio of the maximum diameter to the minimum diameter of the optic disc was >1.2, the discs were subdivided into vertically oval, with the major axis ±10 degrees of the vertical meridian; obliquely oval, with the major axis between 11 and 69 degrees; and horizontally oval, with the major axis ±20 degrees of the horizontal meridian (Fig 1F and G).

Statistical Analyses

Age, refractive error, axial length, best-corrected visual acuity in logarithm of the minimum angle of resolution units, choroidal thickness, and scleral thickness were compared between the 2 groups using Mann–Whitney *U* tests. The incidence of various macular/disc lesions was compared between groups using chi-square tests or Fisher exact probability tests. To determine the statistical significance of deviations in macular/disc lesions from the theoretically expected distribution of the 2 groups (with and without DSM), a binomial test also was performed. To identify the specific factors contributing to the presence of DSM, multivariable logistic regression analyses were performed. Multivariable logistic regression analyses were performed to adjust for age and axial length to specifically evaluate the contribution of DSM to the presence of any macular/disc lesions and to each macular/disc lesion. Multivariable logistic regression analyses also were performed to identify the influence of age, axial length, and presence of DSM on subfoveal choroidal thickness. A P value <0.05 was considered statistically significant. When both eyes of the same individuals were included, the right eye was chosen for statistical analyses.

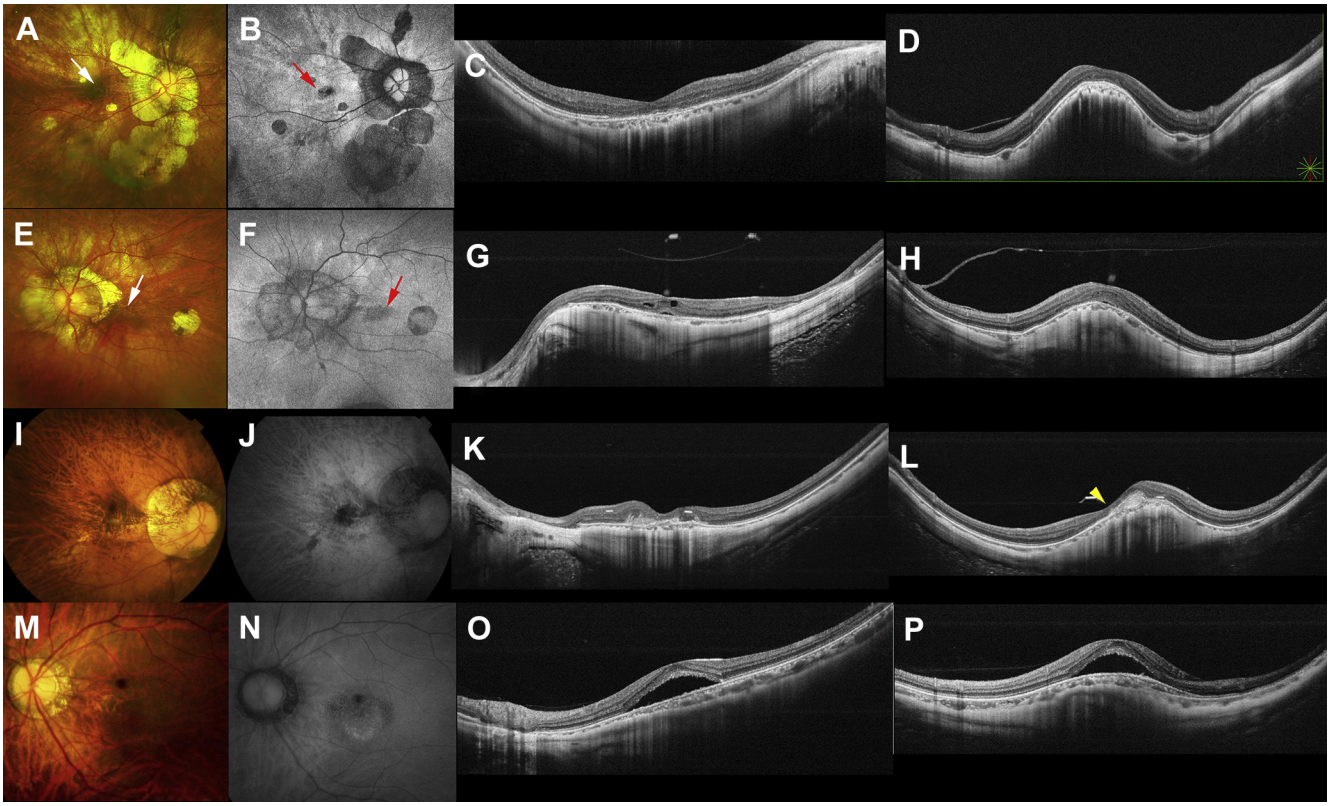

Figure 3. A dome-shaped macula (DSM) seen only in the vertical optical coherence tomography (OCT) section through the fovea (horizontally oriented DSM). **A,** Right fundus of a 63-year-old woman with an axial length of 31.4 mm. Macula shows increased pigmentation (*arrow*). **B,** In the fundus autofluorescence (FAF) image, the macular pigmentation is observed as hypoautofluorescence (*arrow*). **C, D,** Optical coherence tomography images across the central fovea. An inward bulge due to a DSM is observed in the vertical OCT section **(D)** but not in the horizontal section **(C)**. The DSM shows an abrupt and high bulge. **E,** Left fundus of a 52-year-old man with an axial length of 31.1 mm. Macula has increased pigmentation (*arrow*). **F,** Fundus autofluorescence image shows hypoautofluorescence in the macula (*arrow*). **G, H,** Optical coherence tomography images across the central fovea. An inward bulge due to a DSM is observed in the vertical OCT section **(H)** but not in the horizontal section **(G)**. **I,** Right fundus of a 47-year-old woman with an axial length of 29.5 mm. This patient has a Fuchs' spot due to myopic choroidal neovascularization (CNV). **J,** Fundus autofluorescence image shows hypoautofluorescence in the macula due to the Fuchs' spot. **K, L,** Optical coherence tomography images across the central fovea. An inward bulge due to a DSM is observed only in the vertical OCT section **(L)** and not in the horizontal section **(K)**. Scarred CNV is also observed. The central fovea (*arrowhead* in **L**) is located superior to the top of the dome. Although an inward bulge is not seen in the horizontal OCT section, the scleral thickness is increased in the macula and the curvature of the outer surface of the sclera protrudes posteriorly. **M,** Left fundus of a 77-year-old woman with axial length of 30.9 mm. The black dot superior to the center of the photograph is an artifact. **N,** Fundus autofluorescence image shows granular hyperautofluorescence corresponding to subretinal precipitates within the serous retinal detachment (RD). A black dot in the center of the photograph is an artifact. **O, P,** Optical coherence tomography images across the central fovea. An inward bulge due to a DSM is observed only in the vertical OCT section **(P)** and not in the horizontal section **(O)**. Serous RD is observed in the macula.

Results

A total of 543 consecutive patients (1086 eyes) with bilateral high myopia and 43 patients (43 eyes) with unilateral high myopia who had OCT examinations through the central fovea at our High Myopia Clinic between February 2012 and November 2013 were enrolled. Among them, 11 eyes of 11 patients were excluded because the OCT images were not clear enough because of cataract. A total of 1118 highly myopic eyes of 586 patients were included in the present study for retrospective analyses.

Among the 1118 eyes, 225 (20.1%, 160 patients) had a DSM that met the definition.[4] The DSM was bilateral in 65 patients and unilateral in 95 patients. The DSMs were classified into 3 patterns according to the slightly modified method described by Caillaux et al[3]: (1) DSM present in both the vertical and horizontal OCT sections (bidirectional DSM); (2) DSM present in only the vertical section (horizontally oriented DSM); and (3) DSM present in only the horizontal section (vertically oriented DSM). Among the 225 eyes, a bidirectional DSM was found in 46 eyes (20.4%) (Fig 2), a horizontally oriented DSM was found in 174 eyes (77.3%) (Fig 3), and a vertically oriented DSM was found in only 5 eyes (2.2%) (Fig 4).

Comparison of Demographics of Highly Myopic Eyes with and without a Dome-Shaped Macula

The demographics of 225 eyes with DSM and 893 eyes without DSM are summarized in Table 1. Statistical comparison between the highly myopic eyes with and without DSM showed that the highly myopic patients with DSM were significantly younger and more myopic, and had significantly longer axial length and better

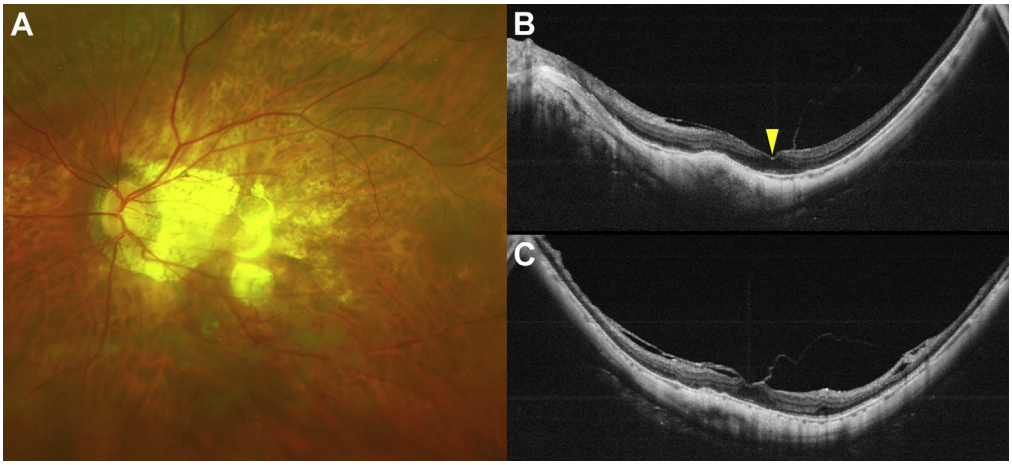

Figure 4. A dome-shaped macula (DSM) seen only in the horizontal optical coherence tomography (OCT) sections through the fovea (vertically oriented DSM). **A,** Left fundus of a 73-year-old woman with axial length of 31.0 mm. **B, C,** Optical coherence tomography images across the central fovea. An inward bulge due to a DSM is observed only in the horizontal OCT section **(B)** and not in the vertical section **(C)**. The central fovea (*arrowhead* in **B**) is located temporal to the top of the dome.

vision. The mean dome height in the 225 eyes was 188.2±111.9 μm (range, 55–634 μm).

Three OCT instruments were used to measure choroidal and scleral thicknesses. The Cirrus OCT instrument was used in 36 of 225 eyes (16.0%) with a DSM and 185 of 893 eyes (20.7%) without a DSM, the RS3000 instrument was used in 69 of 225 eyes (30.7%) with DSM and in 216 of 893 eyes (24.2%) without a DSM, and a swept-source OCT instrument was used in 120 of 225 eyes (53.3%) with a DSM and in 492 of 893 eyes (55.1%) without a DSM. There was no statistical difference in the choroidal and scleral thickness measured by the 3 OCT machines for eyes with and without a DSM.

The subfoveal choroidal thickness was not significantly different between the eyes with DSM and the eyes without a DSM (97.0±37.5 vs. 105.5±41.5 μm, respectively). The results of multivariable logistic regression for subfoveal choroidal thickness showed that age and axial length were significantly associated with choroidal thickness, but the presence of a DSM was not (Table 2, available at www.aaojournal.org). The subfoveal scleral thickness was measured in only 102 of the 225 eyes with a DSM; thus, the subfoveal scleral thickness between the 2 groups was not statistically compared. The scleral thickness 3 mm above and beneath the fovea was measurable in all cases with a DSM and without a DSM. There was no significant difference in the scleral thickness 3 mm superior to and inferior to the fovea between these 2 groups (212.4±79.6 vs. 208.6±48.6 μm and 214.6±77.7 vs. 196.9±45.0 μm, respectively; Table 1). These findings support the data found by Imamura et al.[2]

Presence of a Dome-Shaped Macula in Eyes with Myopic Macular and Disc Complications

Between eyes with and without a DSM, comparisons were made of the incidence of the macular lesions that had been reported to develop in eyes with a DSM.[1–4,7,11,12] First, chi-square or Fisher exact probability tests were performed to determine the incidence

Table 1. Patient and Eye Demographics between Highly Myopic Eyes with and without a Dome-Shaped Macula

Scleral thickness (mm)	DSM		*P* Value
	Present (n = 225 eyes)	*Absent (n = 893 eyes)*	
No. of eyes (patients)	225 (160)	893 (512)	-
Age (yrs)	62.2±11.5 (31–87)	66.0±13.1 (14–91)*	<0.0001
Refractive error (D)†	−15.8±3.9 (−8.0 to −31.0)	−13.3±5.0 (−5.0 to −30.0)	<0.0001
Axial length (mm)	30.7±2.0 (25.9–35.7)	30.0 ±2.1 (24.9–40.0)	<0.0001
logMAR BCVA	0.22±0.35 (−0.18 to 2.0)	0.42±0.55 (−0.18 to 2.0)	0.015
Subfoveal choroidal thickness (mm)	97.0±37.5 (54–198)	105.5±41.5 (55–305)	0.06
Subfoveal scleral thickness (mm)§	380.2±115.6 (162–678) (n = 102 eyes)	258.0±65.0 (101–455) (n = 558 eyes)	-
3 mm above fovea (mm)	212.4±79.6 (43–500)	208.6±48.6 (89–374)	0.347
3 mm below fovea (mm)	214.6±77.7 (36–500)	196.9±45.0 (88–394)	0.134

BCVA = the best-corrected visual acuity; D = diopter; DSM = dome-shaped macula; logMAR; logarithm of the minimum angle of resolution; SD = standard deviation.
Data are mean standard deviation (range) unless otherwise indicated.
*Data of 86 patients with a DSM in the fellow eye were excluded; thus, data from only 426 patients were used for the analysis.
†Data of 456 eyes with a history of cataract surgery were excluded.
§The subfoveal scleral thickness was not measured in the eyes with a too-thick sclera or unclear border of the outer surface of the sclera.

of each macular/disc lesion between highly myopic eyes with and without a DSM. Overall, the rate of macular/disc lesions was significantly higher in eyes with a DSM than in eyes without a DSM. A serous RD was detected significantly more frequently in eyes with a DSM than in eyes without a DSM, although only 6 eyes had a serous RD. Four of 6 eyes with serous RD had a DSM; 2 eyes had a bidirectional DSM, and 2 eyes had horizontally oriented DSMs. The 2 eyes with a serous RD but without a DSM also had a mild macular bulge; however, the maximum dome height was 43 and 39 μm, respectively, which did not meet the definition of a DSM.

Foveal RS was detected significantly more frequent in highly myopic eyes without a DSM than in those with a DSM. By contrast, an extrafoveal RS was detected significantly more frequently in highly myopic eyes with a DSM than in those without a DSM, supporting the data provided by Ellabban et al.[4]

The rate of macular CNVs was detected significantly more frequently in eyes without a DSM than in eyes with a DSM. There was no significant difference in the rate of lamellar macular hole and full-thickness macular hole between the eyes with and without a DSM.

A total of 187 eyes had at least 1 of the 3 funduscopic features, that is, macular pigmentation, appearance of a horizontal ridge connecting the optic disc and the fovea, and a horizontally oval optic disc. Only 1 eye had all 3 features, and 29 eyes had 2 of the 3 features. The remaining 157 eyes had only 1 of these 3 features. All of these 3 features were significantly more frequent in eyes with a DSM than in eyes without a DSM.

The highly myopic patients were examined and followed at our High Myopic Clinic and were not recruited because of the presence of a DSM. Thus, binomial tests were performed to determine the statistical significance of deviations in macular/disc lesions from the theoretically expected distribution of the 2 groups, with a DSM and without a DSM, and to compare the rate of DSMs in eyes with each category of macular/disc lesions (Table 3). Results similar to those obtained by chi-square tests or Fisher exact probability tests were also obtained except in 2 categories: "any kind of complications" and "CNV." In the eyes with CNV, 16.4% had a DSM and 83.6% did not have a DSM; however, there was no significant difference in the distribution of CNVs and the distribution of DSMs.

Among the 3 fundus lesions that we focused on as clues to suspect the presence of a DSM, 64 of the 70 eyes (91.4%) with a horizontal ridge had a DSM. The OCT findings of the horizontal ridge showed a hyporeflective structure within the sclera (Fig 5F) and a notch between the lines (Fig 5G).

The results of multivariable logistic regression for the presence of a DSM and the presence of any macular/disc lesion of each lesion are shown in Table 4. Axial length and age were significantly associated with the presence of a DSM ($P < 0.001$ and $P = 0.043$, respectively). We excluded 2 lesions whose incidence was not different between the eyes with and without a DSM by chi-square tests, Fisher exact probability tests, and binomial tests from further multiple regression analyses. Then the presence of the other 7 lesions (CNV, serous RD, foveal RS, extrafoveal RS, macular pigmentation, appearance of a horizontal

Table 3. Presence and Absence of a Dome-Shaped Macula in Highly Myopic Eyes with Macular and Disc Complications

	Chi-Square Tests or Fisher Exact Probability Tests			Binomial Test		
	DSM			DSM		
	Present	*Absent*	P *Value*	*Present*	*Absent*	P *Value*
Overall (n = 1118 eyes)	225 eyes (20.1%)	893 eyes (79.9%)	NA	225 eyes (20.1%)	893 eyes (79.9%)	NA
No complications (n = 536 eyes)	59/225 eyes (26.2%)	477/893 eyes (53.4%)	<0.001	59/536 eyes (11.0%)	477/536 eyes (89.0%)	<0.001
With any of the following complications (n = 582 eyes)	166/225 eyes (73.8%)	416/893 eyes (46.6%)	<0.001	166/582 eyes (28.5%)	416/582 eyes (71.5%)	<0.001
Macular complication						
CNV (including CNV-related macular atrophy) (n = 293 eyes)	48/225 eyes (21.3%)	245/893 eyes (27.4%)	0.001	48/293 eyes (16.4%)	245/293 eyes (83.6%)	0.062
Serous RD[†] (n = 6 eyes)	4/225 eyes (1.8%)	2/893 eyes (0.2%)	0.017	4/6 eyes (66.7%)	2/6 eyes (33.3%)	0.017
Lamellar macular hole (n = 47 eyes)	8/225 eyes (3.6%)	39/893 eyes (4.4%)	NS	8/47 eyes (17.0%)	39/47 eyes (83.0%)	0.378
Full-thickness macular hole (n = 39 eyes)	7/225 eyes (3.1%)	32/893 eyes (3.6%)	NS	7/39 eyes (17.9%)	32/39 eyes (82.1%)	0.462
Foveal RS (n = 69 eyes)	2/225 eyes (0.9%)	67/893 eyes (7.5%)	<0.0001	2/69 eyes (2.9%)	67/69 eyes (97.1%)	<0.001
Extrafoveal RS (n = 215 eyes)	62/225 eyes (27.6%)	153/893 eyes (17.1%)	<0.0001	62/215 eyes (28.8%)	153/215 eyes (71.2%)	0.001
Other complication						
Macular pigmentation (n = 115 eyes)	58/ 225 eyes (25.8%)	57/893 eyes (6.4%)	<0.0001	58/115 eyes (50.4%)	57/115 eyes (49.6%)	<0.001
Appearance of a horizontal ridge connecting the optic disc and the fovea*[1] (n = 70 eyes)	64/225 eyes (28.4%)	6/893 eyes (0.7%)	<0.0001	64/70 eyes (91.4%)	6/70 eyes (5.2%)	<0.001
Horizontally oval optic disc*[2] (n = 33 eyes)	13/225 eyes (5.8%)	20/893 eyes (2.2%)	0.005	13/33 eyes (39.4%)	20/33 eyes (60.6%)	0.009

CNV = choroidal neovascularization; DSM = dome-shaped macula; NA = not available; NS = not significant; RS = retinoschisis.
*1 and *2; see "Methods" for details of these 2 lesions.
[†]The eyes with CNV were not included in the analyses of the presence of serous RD.

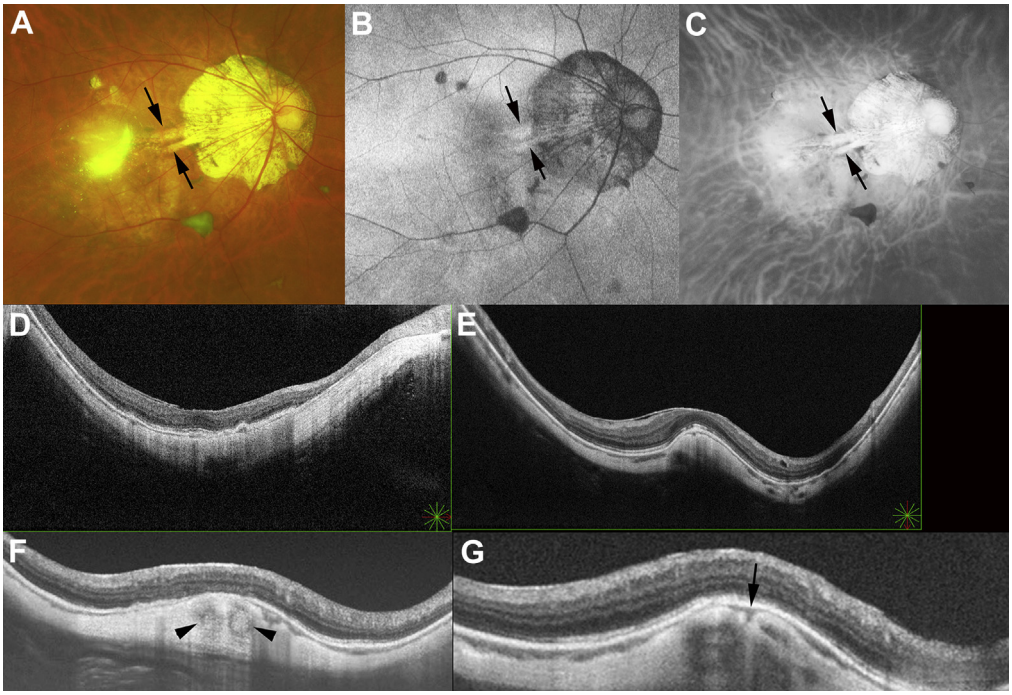

Figure 5. A horizontal ridge running from the optic disc to the macula can be seen in cases with a dome-shaped macula (DSM). **A,** Right fundus of a 72-year-old man with an axial length of 30.2 mm, showing 2 yellowish linear lesions connecting the optic disc and the macula horizontally (*arrows*). **B,** Fundus autofluorescence image shows that the horizontal linear lesions are observed as hyperautofluorescence (*arrows*). **C,** Infrared image shows a horizontal ridge as hyperreflectance (*arrows*). **D, E,** Optical coherence tomography (OCT) images across the central fovea. An inward bulge due to a DSM is observed only in the vertical OCT section **(E)** and not in the horizontal section **(D)**. **F, G,** Vertical OCT images across the horizontal ridge nasal to the central fovea. Two cross-sections of hyporeflective structures (*arrowheads*) are observed in the sclera **(F)**. There is a notch-like dent (*arrow*) between the 2 horizontal protrusions **(G)**.

ridge, horizontal oval optic disc) was analyzed. The results showed that the presence of any of these 7 lesions was significantly associated with age and the presence of a DSM ($P < 0.001$ for both).

Each of the other 7 lesions was then analyzed separately. The results showed that the presence of a DSM was significantly associated with all of the 7 lesions except CNV. Age was the only factor that was significantly associated with CNV in highly myopic patients. Foveal RS and horizontal ridge−like appearance were solely associated by the presence of a DSM. Other lesions were affected by more than 1 factor. Among them, serous RD, macular pigmentation, and horizontally oval optic disc were associated with all of the 3 factors (age, axial length, and presence of a DSM).

Discussion

Our results showed that a DSM was found in 225 of 1118 highly myopic eyes (20.1%). This rate is similar to that reported by Chebil et al,[15] who found a DSM in 24 of 200 highly myopic eyes (12.0%). These results suggest that DSM is a frequent complication in eyes with pathologic myopia. Chebil et al[15] also suggested that DSM might be missed in routine clinical examinations of highly myopic patients unless OCT (especially a vertical scan across the fovea) is performed.

Among the 225 eyes, 77.3% had a horizontally oriented DSM and 20.0% had a bidirectional DSM. This confirmed earlier studies showing that a horizontally oriented DSM was by far the most common type of DSM,[3,4] although there is some inconsistency among earlier studies whether only bidirectional DSMs were included as DSMs[1,2] or whether horizontally oriented DSMs were also included.[3,4,7] Vertically oriented DSMs were seen in only 5 eyes. The vertical ridge in Curtin's type IX staphyloma should be differentiated from vertically oriented DSMs, especially when the ridge exists close to the optic disc.

The patients with a DSM were significantly younger and had significantly longer axial lengths than highly myopic patients without a DSM. The results of multiple regression analyses showed that both age and axial length were significantly associated with the presence of a DSM. Three-dimensional magnetic resonance imaging analyses of the globe recently showed that more than one half of the eyes with a DSM did not have a staphyloma; the eyes were simply elongated into a barrel-shaped globe.[22] This suggested that the DSM group included more patients with a barrel-shaped globe and younger patients who had not developed a staphyloma. However, further investigations are necessary to clarify the reason for the differences in the demographics.

There was no significant difference in the subfoveal choroidal thickness between the eyes with and without a DSM. The results of multiple regression analyses showed that age and axial length were significantly associated with the subfoveal choroidal thickness in highly myopic eyes, as

Table 4. Multivariable Logistic Regression Analysis for the Presence of a Dome-Shaped Macula and its Complications

	Presence of DSM	Complications: Presence of at Least 1 Complication*	CNV (Including CNV-Related Macular Atrophy)	Serous RD†	Foveal RS	Extrafoveal RS	Macular Pigmentation‡	Appearance of a Horizontal Ridge Connecting the Optic Disc and the Fovea‡	Horizontally Oval Optic Disc‡
Adjusted OR									
Age	0.99	1.04	1.03	0.91	1.02	1.02	1.04	1.0068	0.95
Axial length	1.17	0.99	0.99	0.46	1.03	0.96	1.13	1.03	0.7
DSM	–	3.42	0.81	35.5	0.06	1.6	4.98	46.83	3.5
95% CI									
Age	0.98–1	1.02–1.05	1.02–1.05	0.85–0.98	1–1.04	1.01–1.04	1.02–1.06	0.9794–1.035	0.93–0.98
Axial length	1.09–1.2	0.93–1.05	0.92–1.06	0.27–0.79	0.91–1.17	0.89–1.04	1.02–1.26	0.88–1.2	0.58–0.85
DSM	–	2.41–4.86	0.55–1.19	3.05–412.68	0.01–0.47	1.08–2.35	3.17–7.81	19.52–112.35	1.66–7.39
P value									
Age	0.043	<0.001	<0.001	0.011	0.066	<0.001	<0.001	0.629	<0.001
Axial length	<0.001	0.743	0.743	0.002	0.606	0.359	0.021	0.736	<0.001
DSM	–	<0.001	0.279	<0.001	<0.001	0.02	<0.001	<0.001	0.002

CI = confidence interval; CNV = choroidal neovascularization; DSM = dome-shaped macula; OR = odds ratio; RD = retinal detachment; RS = retinoschisis.
*At least 1 of the 7 complications (CNV, serous RD, foveal RS, extrafoveal RS, macular pigmentation, horizontal ridge–like appearance, horizontally oval optic disc).
†The eyes with CNV were not included in the analyses of the presence of serous RD.
‡See "Methods" for details of macular pigmentation, horizontal ridge–like appearance, and horizontally oval optic disc.

reported by Fujiwara et al,[23] but the presence of a DSM was not significantly associated. These results differ from those of Chebil et al,[15] who reported a thicker subfoveal choroid in the eyes with a DSM than in the eyes without a DSM. The reason for this discrepancy is not clear; however, it is possible that more extremely myopic patients were studied in the current study, and Chebil et al studied eyes with higher domes (Table 5, available at www.aaojournal.org). There was no significant difference in the subfoveal scleral thickness 3 mm superior and inferior to the fovea between the eyes with an without a DSM, which confirmed the results of Imamura et al.[2]

Various kinds of macular lesions have been reported to develop in eyes with a DSM.[1–4,6] However, because of a lack of studies comparing the rate of complications between highly myopic eyes with and without a DSM, it is not clear whether these complications are unique to a DSM or a general feature of eyes with pathologic myopia regardless of the presence of a DSM. Our multiple regression analyses showed that the rates of foveal and extrafoveal RS and serous RD were significantly associated with the presence of a DSM, although serous RD was a rare complication (6 eyes in total). In the 6 eyes with a serous RD, 4 had a DSM and 2 did not. However, the 2 eyes with a serous RD but without a DSM had a mild elevation of the macular sclera, although the dome height did not meet the definition of a DSM. This might suggest that some bulge of the sclera is still important for developing serous RD regardless of whether the bulge is high or low. The discrepancy in the rate of serous RD among studies (Table 5, available at www.aaojournal.org) is not clear; however, the mean dome height was much higher in the study by Caillaux et al,[3] and the difference in the dome height could be one of the causes of serous RD.

Macular CNVs have been reported to be a frequent complication of DSMs (12.2%,[7] 25.0%,[8] 41.2%,[4] and 47.8%[2]) in earlier studies. However, the results of multivariate analyses showed that the overall rate of macular CNV, including all phases of CNV, was significantly associated with the age of, but not with the presence of a DSM in, our patients. The reason for the discrepancy in our study and earlier studies is not clear. However, none of the studies reporting the rate of macular CNV in eyes with a DSM compared highly myopic eyes with and without a DSM. Although even patients without visual symptoms visited our High Myopia Clinic, it is possible that the patients with more severe visual symptoms visited retina specialists in the earlier studies.

A DSM is a pathology that is diagnosed on the basis of OCT findings. However, to identify clues to suspect the presence of a DSM from the fundus appearance, we focused on 3 pathologies: macular pigmentation, appearance of a horizontal ridge connecting the optic disc and the fovea, and a horizontally oval optic disc based on our clinical impression. Among these 3 lesions, the macular pigmentation and horizontal ridge were found significantly more frequently in the eyes with a DSM than in the eyes without a DSM. The horizontal ridge was almost exclusively found in the eyes with a DSM, suggesting that this lesion should be considered an important predictive clue for the presence of a DSM.

The pathogenesis of the horizontal ridge—like appearance is not clear; however, one hypothesis is that some directional force pulled toward the dome top might cause the formation of a horizontal ridge connecting the optic disc and the dome top. The OCT findings showed some structural changes deep in the sclera corresponding to the horizontal line (Fig 5F), suggesting this change was not a superficial change of the sclera. The pathogenesis of macular pigmentation is also uncertain, but the chronic RPE damage at the top of the dome might cause an increase in the pigmentation.

Study Limitations

Patients were those who visited the High Myopia Clinic at the Tokyo Medical and Dental University. Thus, it is possible that the results obtained do not reflect the general myopic population. However, it seems that patients with visual symptoms were more actively enrolled in earlier studies,[1,3,4,7,12] which might explain a high percentage of serous RD and CNV. On the other hand, a large population of highly myopic patients in our High Myopia Clinic has been regularly followed regardless of the presence of visual symptoms. Another limitation is that all of the patients were Japanese and did not include other ethnicities, and all of them had high myopia. Because earlier studies showed that mild myopia or even emmetropic eyes can have a DSM,[1,3,6] this enrollment method might have caused a selection bias. Although OCT is performed routinely for highly myopic patients in our clinic, the possibility that the patients with CNV tended to undergo more OCT examinations cannot be completely eliminated. Earlier studies reported differences in the measurement of retinal thickness[24] or choroidal thickness[25] between swept-source OCT and other spectral-domain OCT devices. By contrast, other studies showed that the choroidal thickness measurement was comparable between swept-source OCT and other spectral-domain OCT instruments.[26,27] It is not certain whether this is applicable to highly myopic eyes; however, we should carefully interpret the data because 3 different OCT instruments were used in this study. Also, we admit that the overall rate of funduscopic features to suspect the presence of a DSM (i.e., macular pigmentation, horizontal ridge, horizontally oval optic disc) was not high.

Despite these limitations, the strength of the present study lies in the large number of highly myopic patients who were examined. Through the examination of a large number of consecutive highly myopic patients, we showed that a DSM was found in 20.1% of highly myopic eyes. A comparison of highly myopic patients with and without a DSM showed that serous RD was a rare complication of DSM and that the rate of CNV was not different between the 2 groups. The appearance of a horizontal ridge connecting the optic disc and the fovea and the existence of macular pigmentation in fundus photographs may be important clues for a DSM being present.

Acknowledgments. The authors thank Professor Emeritus Duco Hamasaki for the critical revision of our article.

References

1. Gaucher D, Erginay A, Lecleire-Collet A, et al. Dome-shaped macula in eyes with myopic posterior staphyloma. Am J Ophthalmol 2008;145:909–14.
2. Imamura Y, Iida T, Maruko I, et al. Enhanced depth imaging optical coherence tomography of the sclera in dome-shaped macula. Am J Ophthalmol 2011;151:297–302.
3. Caillaux V, Gaucher D, Gualino V, et al. Morphologic characterization of dome-shaped macula in myopic eyes with serous macular detachment. Am J Ophthalmol 2013;156:958–67.
4. Ellabban AA, Tsujikawa A, Matsumoto A, et al. Three-dimensional tomographic features of dome-shaped macula by swept-source optical coherence tomography. Am J Ophthalmol 2012;3:578−8.
5. Chinskey ND, Johnson MW. Treatment of subretinal fluid associated with dome-shaped macula. Ophthalmic Surg Lasers Imaging Retina 2013;44:593–5.
6. Errera MH, Michaelides M, Keane P, et al. The extended clinical phenotype of dome-shaped macula. Graefes Arch Clin Exp Ophthalmol 2014;252:499–508.
7. Ohsugi H, Ikuno Y, Oshima K, et al. Morphologic characteristics of macular complications of a dome-shaped macula determined by swept-source optical coherence tomography. Am J Ophthalmol 2014;158:162–170.e1.
8. Viola F, Dell'Arti L, Benatti E, et al. Choroidal findings in dome-shaped macula in highly myopic eyes: a longitudinal study. Am J Ophthalmol 2015;159:44–52.
9. Dirani A, Matet A, Beydoun T, et al. Resolution of foveal detachment in dome-shaped macula after treatment by spironolactone: report of two cases and mini-review of the literature. Clin Ophthalmol 2014;8:999–1002.
10. Shin MK, Byon IS, Park SW, Lee JE. Serous retinal detachment accompanied by MEWDS in a myopic patient with dome-shaped macula. Ophthalmic Surg Lasers Imaging Retina 2014;45:253–5.
11. Tamura N, Sakai T, Tsuneoka H. Spontaneous resolution of foveal detachment in dome-shaped macula observed by spectral domain optical coherence tomography. Clin Ophthalmol 2014;8:83–6.
12. Coco RM, Sanabria MR, Alegria J. Pathology associated with optical coherence tomography macular bending due to either dome-shaped macula or inferior staphyloma in myopic patients. Ophthalmologica 2012;228:7–12.
13. Ellabban AA, Tsujikawa A, Muraoka Y, et al. Dome-shaped macular configuration: longitudinal changes in the sclera and choroid by swept-source optical coherence tomography over two years. Am J Ophthalmol 2014;158:1062–70.
14. Battaglia Parodi M, Zucchiatti I, Fasce F, et al. Dome-shaped macula associated with Best vitelliform macular dystrophy. Eur J Ophthalmol 2015;25:180–1.
15. Chebil A, Ben Achour B, Chaker N, et al. [Choroidal thickness assessment with SD-OCT in high myopia with dome-shaped macula]. J Fr Ophtalmol 2014;37:237–41.
16. Cheour M, Ben Aleya N, Brour J, et al. [Dome-shaped macula: appearance on ultrasound and optical coherence tomography]. J Fr Ophtalmol 2013;36:e145–9.
17. Margolis R, Spaide RF. A pilot study of enhanced depth imaging optical coherence tomography of the choroid in normal eyes. Am J Ophthalmol 2009;147:811–5.
18. Tokoro T, ed. Atlas of Posterior Fundus Changes in Pathologic Myopia. Tokyo: Springer-Verlag; 1998:5–22.

19. Yoshida T, Ohno-Matsui K, Yasuzumi K, et al. Myopic choroidal neovascularization: a 10-year follow-up. Ophthalmology 2003;110:1297–305.
20. Hayashi K, Ohno-Matsui K, Shimada N, et al. Long-term pattern of progression of myopic maculopathy: a natural history study. Ophthalmology 2010;117:1595–611.
21. Ohno-Matsui K, Shimada N, Yasuzumi K, et al. Long-term development of significant visual field defects in highly myopic eyes. Am J Ophthalmol 2011;152:256–65.
22. Ohno-Matsui K. Proposed classification of posterior staphylomas based on analyses of eye shape by three-dimensional magnetic resonance imaging. Ophthalmology 2014;121:1798–809.
23. Fujiwara T, Imamura Y, Margolis R, et al. Enhanced depth imaging optical coherence tomography of the choroid in highly myopic eyes. Am J Ophthalmol 2009;148:445–50.
24. Tan CS, Chan JC, Cheong KX, et al. Comparison of retinal thicknesses measured using swept-source and spectral-domain optical coherence tomography devices. Ophthalmic Surg Lasers Imaging Retina 2015;46:172–9.
25. Matsuo Y, Sakamoto T, Yamashita T, et al. Comparisons of choroidal thickness of normal eyes obtained by two different spectral-domain OCT instruments and one swept-source OCT instrument. Invest Ophthalmol Vis Sci 2013;54:7630–6.
26. Tan CS, Ngo WK, Cheong KX. Comparison of choroidal thicknesses using swept source and spectral domain optical coherence tomography in diseased and normal eyes. Br J Ophthalmol 2015;99:354–8.
27. Copete S, Flores-Moreno I, Montero JA, et al. Direct comparison of spectral-domain and swept-source OCT in the measurement of choroidal thickness in normal eyes. Br J Ophthalmol 2014;98:334–8.

Footnotes and Financial Disclosures

Originally received: December 24, 2014.
Final revision: March 15, 2015.
Accepted: April 9, 2015.
Available online: May 16, 2015. Manuscript no. 2014-2089.

[1] Department of Ophthalmology and Visual Science, Tokyo Medical and Dental University, Tokyo, Japan.

[2] Department of Ophthalmology, Cathay General Hospital, Taipei, Taiwan.

Presented in part at the American Academy of Ophthalmology Annual Meeting, October 18−21, 2014, Chicago, Illinois.

Financial Disclosure(s):
The author(s) have no proprietary or commercial interest in any materials discussed in this article.

Supported in part by Research Grant 25670727 from the Japan Society for the Promotion of Science, Tokyo, Japan. The funding organization had no role in the design or conduct of this research.

Author Contributions:

Research design: Liang, Ohno-Matsui

Data acquisition and/or research execution: Liang, Shimada, Tanaka, Nagaoka, Moriyama, Yoshida, Ohno-Matsui

Data analysis and/or interpretation: Liang, Ohno-Matsui

Obtained funding: Not applicable

Manuscript preparation: Liang, Ohno-Matsui

Abbreviations and Acronyms:
CNV = choroidal neovascularization; **DSM** = dome-shaped macula; **FAF** = fundus autofluorescence; **OCT** = optical coherence tomography; **RD** = retinal detachment; **RPE** = retinal pigment epithelium; **RS** = retinoschisis.

Correspondence:
Kyoko Ohno-Matsui, MD, PhD, Department of Ophthalmology and Visual Science, Tokyo Medical and Dental University; 1-5-45 Yushima, Bunkyo-ku, Tokyo 113-8510, Japan. E-mail: k.ohno.oph@tmd.ac.jp.