

Research Article

Spectral-Domain Optical Coherence Tomography-Based Morphofunctional Characterization of Dome-Shaped Maculopathy in Indian Population

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Purpose. To study the clinicodemographic profile of dome-shaped maculopathy (DSM) eyes in the Indian population and characterization using spectral-domain optical coherence tomography (SD-OCT). **Methods.** This observational cross-sectional study included 25 eyes of 14 patients diagnosed with DSM. All eyes underwent SD-OCT for characterization of the dome profile and also to measure central macular thickness (CMT), subfoveal choroidal thickness (SFCT), and dome height (DH) and to detect the presence of subretinal fluid (SRF). **Results.** The mean age of patients was 48.36 ± 14.23 years (range, 28–65 years). Eleven patients had bilateral involvement. Mean axial length of all eyes was 24.25 ± 1.95 mm and mean spherical equivalent -4.23 ± 3.79 DS. Overall, 11/25 eyes (44%) had round domes, 9/25 eyes (36%) had horizontal domes, and 5/25 eyes (20%) had vertical domes, with a mean dome height at fovea of $500.54 \pm 291.58 \mu\text{m}$. Vertical domes had higher DH compared to horizontal or combined domes ($p = 0.02$). Six eyes (6/25, 24%) showed the presence of SRF; 60% of vertical domes had SRF, and 22.2% of horizontal domes had SRF. The eyes having SRF had significantly higher CMT ($p = 0.017$) and DH ($p = 0.001$), especially in horizontal domes ($p = 0.023$). The eyes with thicker SFCT tended to have higher DH and poorer visual acuity. **Conclusion.** Indian DSM eyes may have relatively lesser amounts of myopia. Choroidal thickening may play a role in development of DSM and may also be related to development of subretinal fluid in such eyes.

1. Introduction

Myopia is one of the leading causes of visual morbidity around the world [1, 2], with a particularly high prevalence in the Eastern and the southeastern global population [3–9]. Dome-shaped maculopathy (DSM) is an unusual clinical entity, first described in 2008 by Gaucher and associates as an abnormal forward convex bulge of the macula within concavity of posterior staphyloma in highly myopic eyes [10–12]. Although the prevalence of DSM has not been reported by any population-based study, incidence of DSM in highly myopic eyes has been estimated as 20% in Japan [13], 16% in Turkey [14], and 11% in Europe [15]. DSM has been documented in adults, adolescents, and children in

many other parts of the world, namely, Canada [11], USA [16, 17], UK [18, 19], Europe [10, 15, 20, 21], Korea [22, 23], and Japan [16, 24]. Although, DSM was initially believed to occur exclusively in high myopic eyes with staphyloma, it has also been described in emmetropes, hypermetropes [18], and eyes without staphyloma [16]. The field of retinal imaging by optical coherence tomography (OCT), especially enhanced depth imaging and swept source OCT for deeper penetration into choroid and the sclera, has especially aided in describing the topographic attributes of DSM [16, 24].

After a thorough literature review, we came to the conclusion that no study has been performed to describe the clinical and demographic profile of DSM in the Indian population. Hence, the current study was performed with the

purpose of providing an insight into the clinical picture of dome-shaped maculopathy and relationship between its structural profile and visual function in the Indian population.

2. Methodology

This observational cross-sectional study was performed at a tertiary care ophthalmic center in South India. The study adhered to the Declaration of Helsinki, and ethical clearance was obtained from the Institutional Ethics Committee. Any eye diagnosed as DSM between January 2018 and December 2019 was included for evaluation. Eyes with macular degeneration due to any cause, macular holes, diabetic retinopathy, and significant media opacity obstructing image capturing and patients with subretinal fluid due to any specific retinal conditions such as choroidal neovascularization and diabetic macular edema were excluded.

A complete ophthalmological examination was performed for all subjects, including best corrected visual acuity (BCVA) with Snellen chart and anterior and posterior segment examination and spectral-domain optical coherence tomography (SD-OCT) with enhanced depth imaging (Heidelberg Spectralis, Heidelberg, Germany). All cases of DSM with SRD detected on SD-OCT underwent simultaneous fluorescein angiography and indocyanine green angiography (Heidelberg Spectralis, Heidelberg, Germany) to rule out any underlying choroidal neovascularization. SD-OCT examination was performed in both the eyes of each subject. SD-OCT images were analyzed for measuring the central macular thickness, subfoveal choroidal thickness, dome height, and subretinal fluid height if present. The OCT thickness measurements were all performed between 10am and 2pm during the day for all included patients, to rule out variability of choroidal thickness. Axial length of each eye was measured using partial coherence interferometry (IOLMaster 500, Carl Zeiss Meditec, Jena, Germany).

Dome-shaped maculopathy was defined as a convex elevation of the sclero-choroido-retinal macular complex seen in horizontal and/or vertical SD-OCT scans. Three-dimensional OCT topography was used to identify the orientation of the dome in the posterior pole in addition to the raster scans, and accordingly, types of domes were defined as horizontal oval (when dome was more convex along the vertical axis), vertical oval (when dome was more convex along the horizontal axis), and round (when dome was symmetric along the vertical and horizontal axis). This was performed based on the classification by Caillaux et al. [20]. Dome height (DH) was determined by drawing a line passing through the center of the fovea (line 1), perpendicular to another line (line 2) passing horizontally tangential to the outer border of the line corresponding to the retinal pigment epithelium (RPE) at the edges of the dome [20]. DH was defined as the distance between the intersection of line 1 with RPE and intersection of lines 1 and 2 (Figure 1).

2.1. Statistical Analysis. Data were entered in a Microsoft Excel spreadsheet. Data normality was checked using histograms. Statistical analysis was performed using SPSS for

Windows software (version 20.0, International Business Machines Corp.). Mean (\pm standard deviation) and frequency (percentage) were used to describe continuous and categorical variables, respectively. Quantitative data were compared using the *t* test for parametric and Mann-Whitney *U* test for nonparametric variables. Statistical significance was taken at 2-tailed *p* value of less than 0.05.

3. Results

We analyzed 25 eyes of 14 patients who presented to our center between January 2018 and December 2019 and diagnosed as having dome-shaped maculopathy. The mean age of patients was 48.36 ± 14.23 years (range, 28–65 years). Eleven patients had bilateral involvement (22 eyes). Baseline visual acuity of all eyes was 0.33 ± 0.3 logMAR (Snellen equivalent 6/9). Mean axial length of all eyes was 24.25 ± 1.95 mm (range, 21.4–28.7 mm). The mean spherical equivalent (SE) of 25 eyes was -4.23 ± 3.79 DS (range, +0.50–13 DS). 44% of eyes (11) had myopia less than -3 DS. The mean SE of 22 eyes showing bilateral involvement was -4.65 ± 3.86 DS (median, -3.50 DS). Three eyes showing unilateral involvement had a median SE of -1.25 DS. 23 eyes had myopia, 1 eye had hyperopia, and 1 eye had no significant refractive error. Demographic characteristics of the study population are summarized in Table 1.

The mean dome height at fovea was 500.54 ± 291.58 μ m (range, 116–1311 μ m). None of the eyes exhibited vitreomacular traction, and only 1 eye had an epiretinal membrane. A total of 11/25 eyes (44%) had round domes, 9/25 eyes (36%) had horizontal domes, and 5/25 eyes (20%) had vertical domes. On comparison of clinical characteristics among the three different types of domes (Table 2), we observed that the domes differed significantly in terms of dome height, with vertical domes having higher dome height as compared to horizontal or combined domes ($p = 0.02$). There was no difference in the dome height between horizontal domes and round domes ($p = 0.9$). Rest all clinical parameters among the groups were comparable.

A total of 6/25 eyes (24%) showed the presence of subretinal fluid (SRF) and associated neurosensory detachment. 60% of vertical domes had SRF, and 22.2% of horizontal domes had SRF. Only 1 eye with round dome had SRF. The clinical summary of eyes based on presence of SRF is shown in Table 3.

We observed that eyes having SRF showed significantly higher CMT ($p = 0.017$) and dome height ($p = 0.001$), especially in horizontal domes ($p = 0.023$). The overall dome height was significantly higher in the eyes having SRF, and this difference was significant in horizontal domes, and vertical domes also showed a trend towards higher dome height in the presence of SRF (Table 2). Of the eyes with SRF, 3 eyes were treated with subthreshold micropulse yellow laser and 2 eyes were injected with 1.25 mg/0.05 mL of intravitreal bevacizumab. After a follow-up period of 3 months, the SRF had reduced in all of these eyes, however, still persisted. One eye with SRF was observed, and the SRF persisted till the last follow-up.

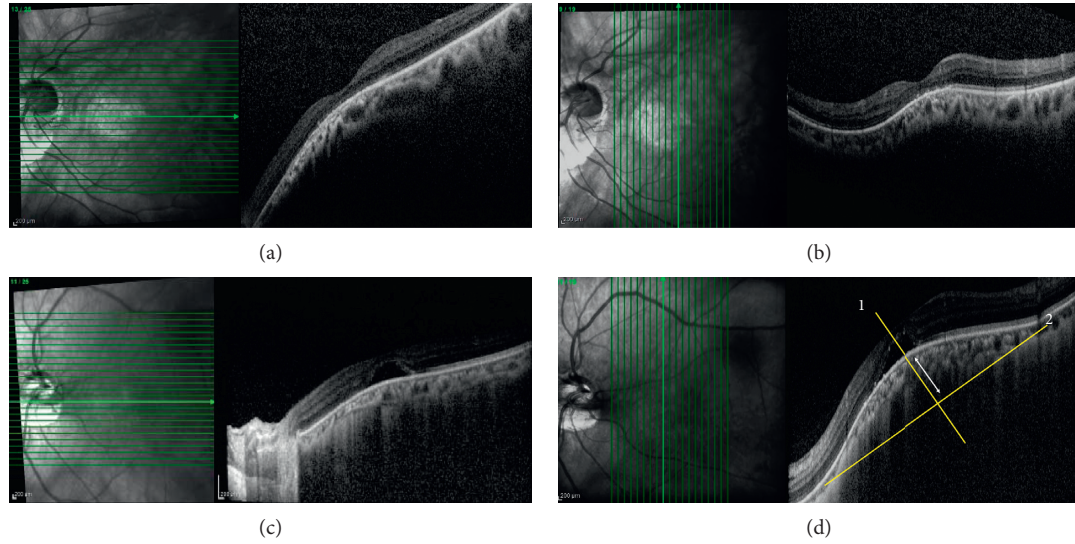


FIGURE 1: Horizontal (a and c) and vertical (b and d) spectral-domain optical coherence tomography (SD-OCT) scans at the fovea of a vertical dome (upper panel) and a horizontal dome with subretinal fluid (SRF) (lower panel). (d) Measurement of dome height in a horizontal dome with SRF on SD-OCT scan—line 1 is a line passing through the center of the fovea, perpendicular to line 2 passing tangential to the outer border of the retinal pigment epithelium (RPE) at the edges of the dome. The distance between the intersection of line 1 with RPE and intersection of lines 1 and 2 represents the dome height.

TABLE 1: Clinical characteristics of study population.

Variable	Mean \pm SD	Range
Number of eyes/number of patients	25/14	—
Sex	8 female/6 male	—
Age (years)	48.36 ± 14.23	28–65
Best corrected visual acuity (logMAR)	0.33 ± 0.30	0–1
Spherical equivalent (DS)	-4.23 ± 3.79	–13.0–0.5
Axial length (mm)	24.25 ± 1.95	21.4–28.7
Subfoveal choroidal thickness (μm)	275.08 ± 82.83	83–422
Central macular thickness (μm)	256.76 ± 108.17	70–455
Dome height (μm)	500.54 ± 291.58	116–1311

TABLE 2: Comparison of parameters among different orientations of myopic domes.

Variable	Horizontal	Vertical	Round	<i>p</i> value
Number of eyes	9	5	11	—
Sex	5 M/1 F	1 M/2 F	5 F	—
Age (years)	42.33 ± 15.98	48 ± 14.93	55.8 ± 10.15	0.318
Best corrected visual acuity (logMAR)	0.35 ± 0.28	0.2 ± 0.44	0.36 ± 0.26	0.198
Spherical equivalent (DS)	-3.78 ± 4.99	-4.2 ± 3.25	-4.61 ± 3.14	0.413
Axial length (mm)	23.75 ± 2.34	24.06 ± 2.06	24.74 ± 1.59	0.312
Subfoveal choroidal thickness (μm)	234.0 ± 97.01	303.6 ± 66.82	295 ± 69.15	0.181
Central macular thickness (μm)	245.2 ± 116.88	318.2 ± 111.86	238.2 ± 98.80	0.627
Dome height (μm)	412.22 ± 247.97	811.2 ± 338.5	424.7 ± 209.49	0.02*
Subretinal fluid (present/absent)	2/7	3/2	1/10	0.095
Foveoschisis (present/absent)	0/9	0/5	2/9	0.670

* Bonferroni post hoc test for dome height: horizontal vs. vertical, -0.03 ; vertical vs. combined, -0.03 ; horizontal vs. combined, -0.9 .

We tried to find an association among different eye characteristics. We found that logMAR visual acuity showed a trend towards a negative association with the subfoveal choroidal thickness (SFCT) ($\rho = 0.307$), indicating that with an increase of choroidal thickness, VA tended

to worsen (Figure 2(a)). Moreover, dome height showed a trend towards a positive association with both SFCT ($\rho = 0.366$) and CMT ($\rho = 0.401$) (Figures 2(b) and 2(c)). The associations were, however, not found statistically significant.

TABLE 3: Impact of SRF on morphological and functional parameters.

Number	Overall SRF			Vertical SRF			Horizontal SRF		
	No 19	Yes 6	<i>p</i>	No 2	Yes 3	<i>p</i>	No 7	Yes 2	<i>p</i>
VA (logMAR)	0.29 ± 0.24	0.44 ± 0.47	0.69	0	0.33 ± 0.58	0.99	0.29 ± 0.17	0.59 ± 0.59	0.67
Spherical equivalent (DS)	-4.41 ± 4.01	-3.67 ± 3.25	0.73	-1 ± 0.35	-6.33 ± 2.02	0.2	-4.71 ± 5.34	-0.5 ± 0.71	0.22
Central macular thickness (μm)	226.58 ± 96.46	352.33 ± 90.79	0.017	242 ± 1.41	369 ± 123.89	0.218	209.71 ± 106.15	369.5 ± 44.55	0.028
Subfoveal choroidal thickness (μm)	264.37 ± 86.61	309 ± 64.12	0.201	299.5 ± 30.41	306.33 ± 91.87	0.914	218.43 ± 105.48	289.5 ± 26.16	0.15
Dome height (μm)	361.16 ± 160.5	900.5 ± 211.96	0.001	495.5 ± 21.92	1021.67 ± 250.64	0.066	309.14 ± 156.28	773 ± 103.23	0.023

VA, visual acuity; SRF, subretinal fluid. * Analysis of round dome was not performed since only 1 eye had presence of SRF. The values in bold are statistically significant at $p < 0.05$.

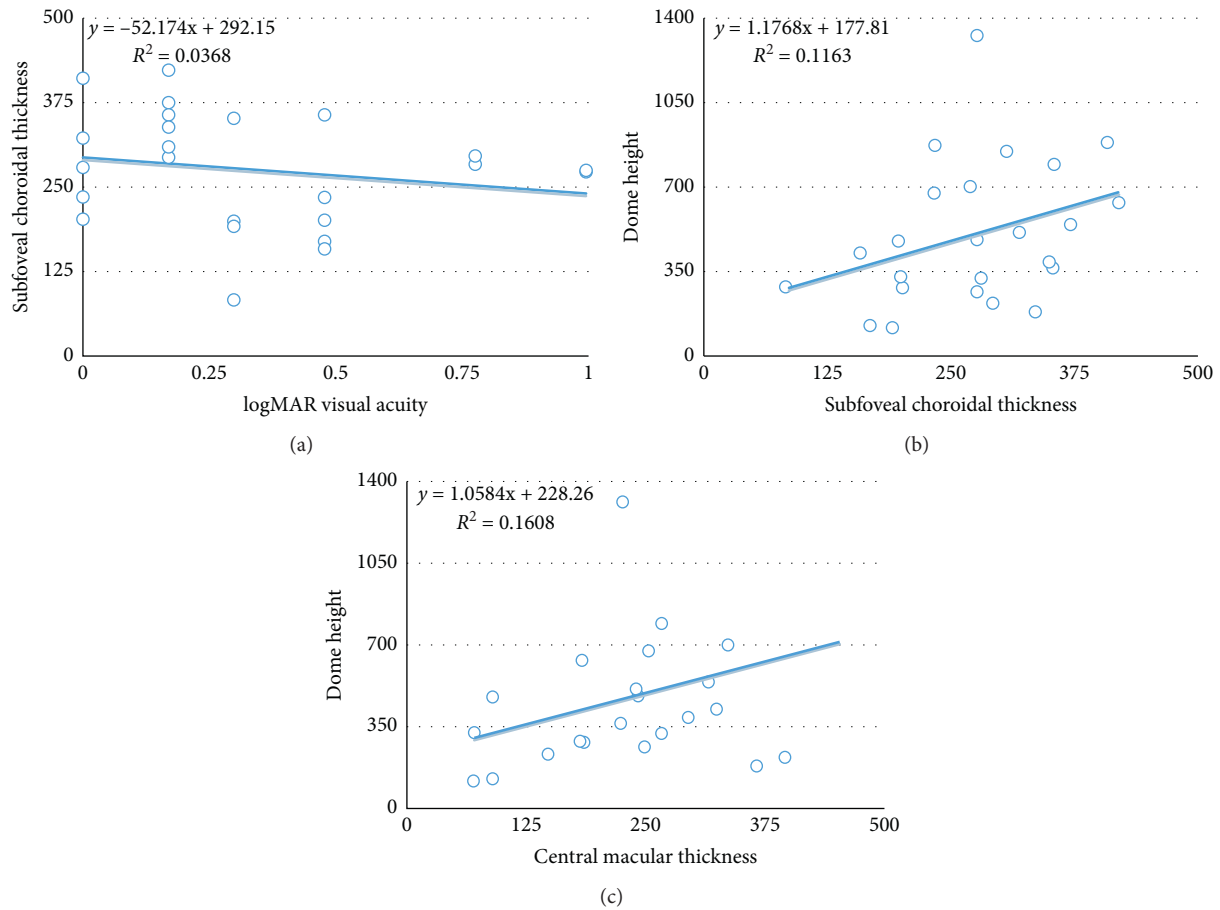


FIGURE 2: (a) Visual acuity (logMAR) showing a negative trend with subfoveal choroidal thickness ($\rho = 0.307$). (b)-(c) Dome height showing a positive trend with SFCT ($\rho = 0.366$) and CMT ($\rho = 0.401$).

4. Discussion

In this study, we have documented the clinical characteristics of Indian eyes with DSM. A noteworthy proportion of DSM eyes, in our study, is that 44% had myopia less than -3 DS, signifying the fact DSM in Indians may be present in low to moderate myopic eyes as well, which may be a different presentation as has been observed in the

Eastern countries. Previously, authors have reported refractive errors of -13.6 DS, -15.5 DS, and -15.8 DS in Asian eyes [13, 16, 25].

The pathomechanism of DSM remains ambiguous with several theories postulated: rigid alteration in the scleral biomechanics with progressing staphyloma, dynamic vitreomacular traction (VMT), locally confined subfoveal choroidal thickening, scleral infolding through collapse of

the posterior portion of the eyewall, compensatory mechanism to myopic globe expansion, adjustment to minimize defocus at the macula, and hypotony in the area of staphyloma [10, 16, 26–28]. Imamura et al. [16] suggested DSM is linked with a localised variation of subfoveal scleral thickness, while it was confirmed later by Ellabban et al. [29] that DSM in high myopes is indeed associated with parafoveal scleral thinning. Thickened sclera can compress the underlying choroidal vasculature with subsequent alterations in retinal pigment epithelium and eventual atrophy [16]. Probably in Indian eyes, localised macular choroidal thickening seems to play a role, as around 1/4th of domes in our series had underlying SFCT greater than 350 μ m. Our data also show that our population may have DSM at a lesser refractive error, which goes against the concept of DSM development secondary to defocus at the macula [27]. Moreover, normal intraocular pressure in all eyes of our series along with the absence of biomicroscopic/OCT signs of vitreomacular traction contravenes the theories of hypotony, scleral collapse, and tangential VMT behind development of DSM. Hence, the pathogenesis of DSM in the Indian population may be different.

OCT imaging has proven to be pivotal in diagnosis of DSM, as domes can be easily missed on routine fundoscopic examination [10, 30]. Caillaux et al. affirmed the relevance of methodical multidirectional SD-OCT scans with at least one horizontal and one vertical scan to diagnose and optimally image the topographic attributes of DSM [20]. In their report, 21% of eyes had round domes, 16% of eyes had vertically oriented oval dome, and 63% of eyes had horizontally oriented oval dome [20]. In our study, round dome was seen to be the most common DSM morphology (44% of eyes), followed by horizontal dome (36% of eyes) and vertical dome (20% of eyes). Contrary to the previous finding that horizontal domes are the most commonly observed DSM pattern [13, 14, 20], we found them to be less common than round domes in our Indian cohort. This might represent a regional variation, although only studies with larger sample size can confirm this observation. We compared the clinical parameters between the three different morphological dome patterns. There was no difference in BCVA and CMT among the three types of domes. The dome height varied among the groups but was significantly higher in vertical domes. The other clinical characteristics were comparable between the groups.

DSM can be associated with a diverse spectrum of vision-threatening macular complications that include serous retinal detachment (SRD), choroidal neovascularization (CNV), pigment epithelial detachment (PED), retinal pigment epithelium (RPE) atrophy, foveoschisis, macular hole (MH), and lamellar MH (LMH) [13, 16, 20, 24, 26, 30–32]. DSM has been observed in about 18% of eyes with myopic choroidal neovascularization [33]. SRD as a macular complication is reportedly very frequent in eyes with DSM [16, 26, 32, 34]. It is also seen commonly in DSM eyes without choroidal neovascularization [20, 31].

A review of literature suggests that there is a marked variation in the incidence of SRF in DSM eyes among different studies, ranging from 1.8% to 66.7% [10, 13, 20, 24, 26, 30, 34].

In our study, SRF was present in 24% of eyes at baseline. CNVM was ruled out by angiography. Lorenzo et al. [11] observed that a lower magnitude of myopia (less than -6 D) may be an important factor influencing the development of SRF. 64% of our study eyes had myopia less than -6 D. We found contrasting results of refractive error in vertical and horizontal domes with SRF (Table 3). Overall, the eyes with SRF were likely to have a significantly greater dome height. Horizontal domes had a significantly lower dome height compared to the vertical domes; however, horizontal domes with a higher dome height were found to be more likely to be complicated by the presence of SRF. For vertical domes, there was no difference in clinical parameters between the eyes with SRF and the eyes without SRF, except for a higher dome height. In keeping with other reports [20, 34], there was a high proportion of vertical domes with SRF (60%). Contrary to the report by Pilotto et al. [34], SRF was least frequently found in round domes in our study (9%).

The precise mechanism behind accumulation of SRF is uncertain. It may be attributable to higher dome height, as suggested by our study. Some of the hypotheses put forward to explain SRF in DSM include altered dynamics of choroidal blood flow and RPE function consequent to a local subfoveal scleral thickening and a central serous chorioretinopathy-like mechanism, wherein locally confined subfoveal choroidal thickening can further enhance slow fluid leakage across the RPE [16]. In our series, although 24% of eyes showed choroidal thickness greater than 350 μ m, only 2 of these eyes had SRF at baseline; hence, it would be difficult for us to exactly establish the relationship between SRF and choroidal thickness.

The various treatment approaches pursued to resolve DSM-related SRF with variable results include observation, anti-VEGF, photodynamic therapy, subthreshold micropulse laser, and mineralocorticoid receptor antagonists [11, 19, 21, 35–37]. A combined treatment approach has been proposed to target both choroidal and RPE dysfunction using half-fluence and half-dose PDT followed by subthreshold 577 nm micropulse laser therapy, wherein SRF diminished in all cases and had completely resolved in 45.4% of the eyes at 6 months [38]. In our series, 5/6 eyes with SRF were given some form of treatment; however, SRF still persisted at the final follow-up of 3 months.

Interestingly, our results also suggest that SRF is not likely to influence visual acuity. There have been contrasting reports regarding outcome of SRF on visual acuity [11, 20, 21, 29, 31, 32, 34, 36, 39, 40]. It has been postulated that shallow serous retinal detachments in DSM eyes might aid sustenance of photoreceptors by ensuring adequate oxygen and nutrient diffusion from the choriocapillaris to the photoreceptors [36]. When the presence of SRF did not significantly impact the vision compared to eyes without SRF in our series, any attempt to reduce SRF by treatment can be questionable. However, only longer periods of follow-up may confirm these findings in a real-world scenario.

5. Conclusions

In summary, dome-shaped maculopathy in Indian population may be seen in eyes with relatively less myopia than

seen globally. Optical coherence tomography helps us in not only characterizing DSM but also in identifying specific features which may be associated with the prognosis of these eyes like the dome height. Choroidal thickening may play a role in development of DSM and may also be related to development of subretinal fluid in such eyes.

Data Availability

The data used to support the findings of this study are available from Dr. Sagnik Sen and Dr. Prithviraj Udaya upon request.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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