

# Evolution of Dome-shaped Macula Is Due to Differential Elongation of the Eye Predominant in the Peri-dome Region



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- **PURPOSE:** To determine the mechanism behind macular bulge height increase in eyes with dome-shaped macula (DSM).
- **DESIGN:** Retrospective, observational case series.
- **METHODS:** Eyes presenting with DSM followed up for a minimum of 1 year were examined using ocular biometry and spectral-domain optical coherence tomography at baseline and at end of follow-up. Axial length (AL), DSM bulge height, and central and peripheral choroidal thickness (nasal, temporal, superior, and inferior quadrants) were reported. Eyes were categorized into 2 groups for comparison: the “mini-DSM” group (DSM < 100  $\mu\text{m}$ ) and the “classic” DSM group (DSM > 100  $\mu\text{m}$ ).
- **RESULTS:** Fifty-eight eyes (33 patients) were studied: 32 (55%) were classic DSM and 26 (45%) mini-DSM. During the mean follow-up of  $51.76 \pm 36.01$  months, mean AL increased from  $26.99 \pm 2.94$  mm to  $27.12 \pm 3.09$  mm ( $P = .010$ ) and mean macular bulge height increased from  $235.88 \pm 282.47$   $\mu\text{m}$  to  $262.34 \pm 317.15$   $\mu\text{m}$  ( $P < .001$ ). DSM height change was significantly higher than AL change ( $P < .001$ ). Mean peripheral choroidal thickness significantly decreased nasally ( $P = .008$ ), temporally ( $P = .026$ ), and inferiorly ( $P < .001$ ). Mini-DSM eyes exhibited shorter AL ( $26.17$  vs  $27.66$  mm;  $P = .027$ ), greater visual acuity ( $0.169$  vs  $0.437$  logMAR;  $P = .002$ ), and fewer macular complications compared to classic DSM eyes.
- **CONCLUSIONS:** Macular bulge increase in DSM is associated with eye elongation and overall thinning of the peripheral choroid. DSM might result from differential elongation of the eye predominant in the peri-dome region. Mini-DSM (ie, inferior to 100  $\mu\text{m}$ ) are characterized by slower evolution, better visual prognosis, and

fewer complications compared to “classic” DSM. (Am J Ophthalmol 2021;224:18–29. © 2020 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

**D**OME-SHAPED MACULA (DSM) WAS RECENTLY described in myopic patients as a macular bulge within the concavity of a posterior staphyloma.<sup>1</sup> This protrusion affects 10.7% to 14.6% of highly myopic eyes.<sup>1–3</sup> Some recent studies have also described this entity in emmetropic and hyperopic eyes without staphyloma.<sup>4,5</sup> DSM is a progressive condition, with the macular bulge height increasing over time.<sup>6,7</sup> The mechanisms underlying this evolution are still unclear and several hypotheses have so far been advanced. Imamura and associates proposed that the perimacular area expands while the macular region remains stable.<sup>4</sup> Others reported perimacular scleral and choroidal thinning, which may support this hypothesis.<sup>4,7,8</sup> Conversely, Fang and associates<sup>9</sup> reported that DSM evolution was associated with the presence of macular Bruch membrane defects (MBMDs). MBMDs can provoke focal relaxation of the posterior sclera, potentially enabling it to partially bulge inward, thus increasing DSM bulge height.<sup>9</sup>

Different types of DSM have previously been described: round, vertical, and horizontal DSM,<sup>6,10</sup> along with more recent description of ridge-shaped macula (RSM).<sup>11</sup> The height of the macular bulge in DSM can vary greatly from a few microns to nearly 1 mm. The differences in form and height of DSM seem to be unevenly associated with vision-threatening conditions. DSM with high bulges seem to present with more severe pigmented epithelium atrophic changes and are commonly associated with serous retinal detachment (SRD).<sup>6</sup> Increasing macular bulge height over time has been previously observed in DSM with relatively high bulges.<sup>6,7</sup> However, it remains unknown whether smaller bulges demonstrate the same evolution, as most studies on the subject only reported DSM with bulges far greater than 100  $\mu\text{m}$  high.<sup>7,11–15</sup>

The main objective of this study was to determine if the increase in DSM height was due to an inward evolution of the bulge or to an outwardly oriented expansion of the perimacular regions. To determine this, changes in axial length

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Accepted for publication Nov 20, 2020.

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(AL) and height of the macular bulge were retrospectively studied in a cohort of DSM patients, also including patients presenting with particularly small DSM.

## METHODS

• **STUDY POPULATION:** This retrospective study included patients with DSM who had undergone a minimal follow-up of 12 months. Data were collected between May 2016 and November 2019 from the Strasbourg University Hospital (Strasbourg, France) and Lariboisière Hospital (Paris, France). All included patients had undergone at least 2 clinical evaluations, including best-corrected visual acuity (BCVA), slit-lamp examination, fundus biomicroscopy, spectral-domain optical coherence tomography (SDOCT), and AL measurement at the beginning and end of follow-up. Diagnosis of DSM was assessed on SDOCT by the presence of a convex elevation of the macula not attributable to another retinal or choroidal disease. Considering that DSM has been previously reported in emmetropic and hyperopic eyes,<sup>5</sup> the emmetropic or hyperopic state and the absence of posterior staphyloma were not considered as exclusion criteria. In the literature, some studies have excluded DSM inferior to 50  $\mu\text{m}$ ,<sup>7,11–15</sup> while some other did not.<sup>1,4–6,8</sup> In the present work, the DSM height inferior to 50  $\mu\text{m}$  was not an exclusion criterion.

All included patients had signed written informed consent for the use of their data in this study. Patients presenting with inferior staphyloma (type V staphyloma, according to the Curtin classification), alone or associated with a tilted disk, were excluded.<sup>16</sup> Eyes with media opacities preventing quality imaging, history of retrofoveal choroidal neovascularization (CNV), or any macular disease other than DSM (except commonly associated complications such as lamellar macular hole or parafoveal schisis) were also excluded. The procedures used in this study were approved by the ethics committee of Strasbourg Medical School and University Hospitals and adhered to the tenets of the Declaration of Helsinki.

• **AXIAL LENGTH ASSESSMENT:** AL was measured by ocular biometry, using partial optical coherence interferometry devices OA-2000 (Tomey GmbH, Nagoya, Japan) and IOL-Master 500 and 700 (Carl Zeiss Meditec, Jena, Germany). All these devices were set to a measurement resolution of 0.01 mm.

• **OPTICAL COHERENCE TOMOGRAPHY MEASUREMENTS:** All measurements were performed using the follow-up tool of the Spectralis HRA + OCT (Heidelberg Engineering, Heidelberg, Germany). Enhanced-depth imaging OCT was performed on both eyes of each patient. A 30-degree horizontal and vertical line setup was used and the

scans were centered on the fovea and automatically averaged (ART setup at 100 scans/line) before recording. OCT measurements were retrospectively performed by 2 investigators in 2 rounds and 3D reconstructions were built when dense multiple-layer scans were available.

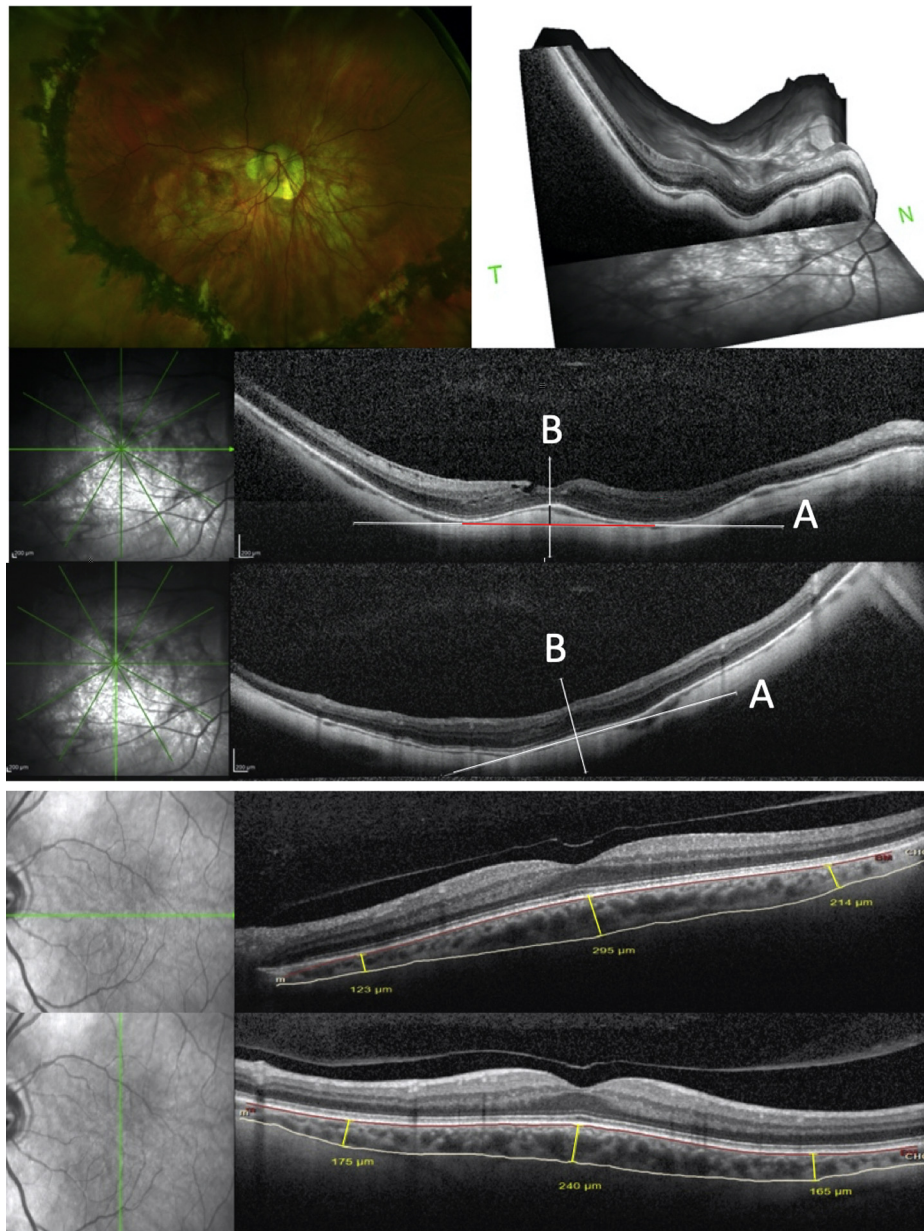
*Macular bulge evaluation.* The macular bulge height was measured on horizontal and vertical SDOCT scans using the Spectralis caliper tool, as previously described.<sup>10</sup> A first line (A) tangent to the outer border of the retinal pigment epithelium was traced. Then, a second line (B), passing through the center of the fovea and perpendicular to the first, was drawn. The bulge height was defined as the distance between the intersection of line A with the outer border of the retinal pigment epithelium and the intersection of line A with line B, as shown in Figure 1. The width of the bulge was also determined for each patient, as described by Xu and associates,<sup>11</sup> measuring the distance between the 2 ends of the lowest points around the macular elevation, along line B (Figure 1).

DSM type was defined according the classification of Caillaux and associates<sup>10</sup>: vertical oval-shaped dome (vertical DSM) (Figures 1 and 2); horizontal oval-shaped dome (horizontal DSM) or round dome (round DSM) (Figure 2). Presence of SRD and central retinal atrophy were noted. Central retinal atrophy was confirmed by autofluorescence imaging (Figure 2) and absence of CNV in patients with SRD was confirmed by fluorescein angiography.

*Choroidal thickness assessments.* Choroidal thickness (CTh) was assessed by the Spectralis caliper tool, manually measuring the distance between the Bruch membrane and the choroidoscleral interface on horizontal and vertical scans. Central CTh was measured at the center of the fovea. Superior, inferior, nasal, and temporal CTh were measured 3000  $\mu\text{m}$  in the superior, inferior, nasal, and temporal directions from the center of the fovea, respectively (Figure 1, Bottom).

• **TYPE OF DOME-SHAPED MACULA:** All analyzed eyes were categorized into 2 different groups: eyes with a bulge height of less than 100  $\mu\text{m}$  into the “mini-DSM” group (Figure 1, bottom images and Figure 2, bottom images) and those with a bulge height greater than 100  $\mu\text{m}$  into the “classic” DSM group (Figure 1, top and middle images and Figure 2, top and middle images). Baseline characteristics of eyes and patients were compared between the groups (ie, visual acuity, macular complications, refractive error (RE), AL, macular bulge height and width, type of DSM, central and peripheral CTh). Evolutions of those parameters were also compared between groups.

• **STATISTICAL ANALYSIS:** The Gaussian distribution of continuous variables was tested by Shapiro-Wilk test. Comparisons of AL, macular bulge height and width, and CTh (central and peripheral) within the follow-up period were performed using parametric tests when Gaussian



**FIGURE 1.** Examples of optical coherence tomography (OCT) measurements. The 4 images on top correspond to measurements of the bulge height and width in the same patient. Ultra-wide-field retinography (Top left) showing typical highly myopic fundus with preventive laser in the retinal periphery. Three-dimensional OCT (Top right) showing vertical oval-shaped dome (vertical dome-shaped macula [DSM]). The 2 images below show horizontal and vertical OCT sections through the fovea with bulge height measurement. White line A corresponds to the tangent to the outer border of the retinal pigment epithelium (RPE) and white line B passes through the center of the fovea, perpendicular to line A. The black line on the horizontal section represents the macular bulge height, here measuring 164  $\mu\text{m}$ . The red line on the horizontal section represents the macular bulge width, along line B, here measuring 2,484  $\mu\text{m}$ . In this example of vertical DSM, bulge height and width are null in the vertical scan (Bottom). The 2 bottom OCT images show choroidal thickness measurements in the left eye of another patient with mini-DSM: the red line showing the delineation of the Bruch membrane and white line the inner scleral border. Central and peripheral choroidal thickness (each at 3,000  $\mu\text{m}$  from the fovea) were measured using the caliper tool of the horizontal (top) and vertical (bottom) OCT scans. In this example of mini-DSM, the bulge height is 84  $\mu\text{m}$  on the horizontal section and 67  $\mu\text{m}$  on the vertical section.

conditions were respected and nonparametric tests in other conditions. The same statistical analyses were performed in both groups of the study (ie, mini-DSM and classic DSM

groups). Comparisons between the mini-DSM and classic DSM groups were performed using Student *t* test and  $\chi^2$  test, where appropriate.



**FIGURE 2.** Examples of autofluorescence imaging (left) with corresponding optical coherence tomography (OCT) (right) in 3 different eyes with DSM. For each eye, the horizontal OCT section is situated above the vertical OCT section. The first classic dome-shaped macula (DSM) (Top) shows a round dome with a macular bulge height of 244  $\mu\text{m}$  on the horizontal section and of 230  $\mu\text{m}$  on the vertical section. Associated complications are central retinal atrophy (CRA) and serous retinal detachment (SRD). The second classic DSM (Middle) shows a vertical oval-shaped dome with a macular bulge height of 768  $\mu\text{m}$  on the horizontal section, while no bulge is observed on the vertical section. Associated complications are CRA and SRD. The third mini-DSM (Bottom) shows a round dome with a macular bulge height of 25  $\mu\text{m}$  on the horizontal section and of 41  $\mu\text{m}$  on the vertical section. Only mild alteration of the pigmentary epithelium is seen on the autofluorescence imaging.

Statistical analyses were performed using GraphPad InStat 3.10 (GraphPad Software, Inc., CA). In all analyses,  $P$  values  $< .05$  were considered statistically significant.

## RESULTS

THE DATA OF 84 EYES FROM 50 PATIENTS PRESENTING WITH DSM were reviewed. Of these, 26 eyes of 18 patients were

excluded owing to lack of data (ie, missing 1 AL assessment or OCT examination from the 2 needed).

- **BASELINE CHARACTERISTICS OF THE STUDIED EYES:** In the end, 58 eyes of 33 patients were included in the study. Nearly half of the eyes (27/58 eyes) presented with round DSM, 19 (32.8%) with vertical DSM, and 12 (20.7%) with horizontal DSM. There were 26 eyes (45%) of 17 patients categorized into the mini-DSM group, while the

**TABLE 1. Baseline Characteristics of Patients**

Characteristic	Result	P <sup>a</sup>
Eyes, n	58 (100%)	-
Subjects, M/F (%)	15 (45.5%)/18 (54.5%)	-
Mini-DSM eyes, n (%) / classic DSM eyes, n (%)	26 (45%)/32 (55%)	-
Age, years, mean ± SD [range]	64.4 ± 12.8 [32 to 788]	-
Bilateral/unilateral condition, n (%)	52 (89.7%)/6 (10.3%)	-
Refractive error, diopters ± SD [range]	-7.74 ± 7.19 [-20.25 to +3.00]	-
AL, mm ± SD [range]	26.99 ± 2.94 [21.61 to 32.71]	-
Macular bulge height, μm ± SD [range]	235.86 ± 282.44 [28 to 1,419]	-
Type of DSM	-	-
Dome, n (%)	27 (46.6)	-
Vertical, n (%)	19 (32.8)	-
Horizontal, n (%)	12 (20.7)	-
Complications	-	-
SRD, n (%)	11 (19.3)	-
Central retinal atrophy, n (%)	15 (26.3)	-
Central CTh, μm ± SD	147.67 ± 89.81	-
Comparison with central CTh at baseline	-	-
Nasal CTh, μm ± SD	-79.50 ± 40.09	<.001*
Temporal CTh, μm ± SD	-18.82 ± 23.81	.03*
Superior CTh, μm ± SD	-24.78 ± 25.92	.005*
Inferior CTh, μm ± SD	-47.67 ± 24.1	<.001*
Follow-up period, months ± SD [range]	51.76 ± 36.01 [12 to 161]	-

AL = axial length; CTh = choroidal thickness; DSM = dome-shaped macula; F = female; M = male; SD = standard deviation; SRD = serous retinal detachment.

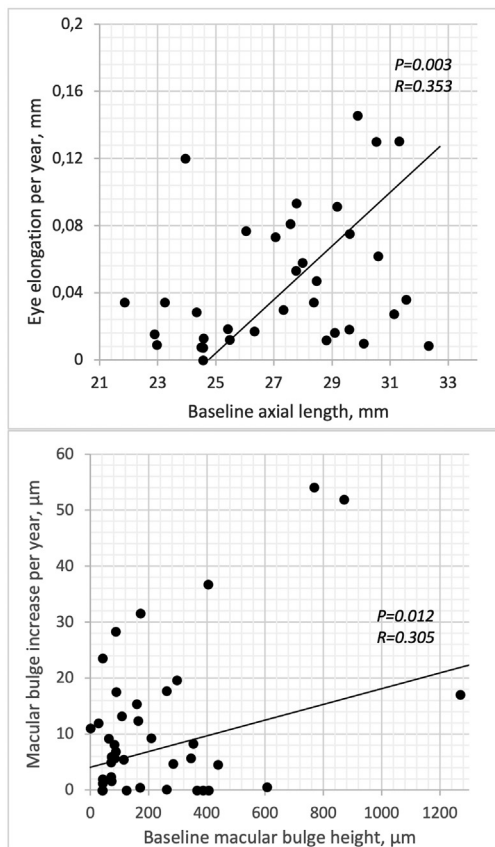
<sup>a</sup>P values for the comparison of peripheral CTh to central CTh at baseline using Student *t* test. (Asterisk indicates statistically significant values.)

**TABLE 2. Mean Visual and Anatomic Changes in the Studied Eyes During Follow-up, and Corresponding Mean Changes Per Year**

	Mean Change During Initial and Final Measurements	P <sup>a</sup>	Corresponding Mean Change per Year
Eyes, n	58 (100%)	-	58 (100%)
AL, mm ± SD	0.13 ± 0.15	.01*	0.04 ± 0.13
Macular bulge height, μm ± SD	26.46 ± 34.68	<.001*	7.39 ± 13.14
Macular bulge width, μm ± SD	-115 ± 18	.32	-51.54 ± 421.51
Central CTh, μm ± SD	-3.2 ± 5.67	.19	-1.06 ± 7.31
Nasal CTh, μm ± SD	-8.02 ± 7.9	.008*	-23.57 ± 26.13
Temporal CTh, μm ± SD	-7.54 ± 10.24	.026*	-1.70 ± 15.34
Superior CTh, μm ± SD	1.88 ± 8.07	.15	-0.88 ± 9.69
Inferior CTh, μm ± SD	-12.73 ± 6.4	<.001*	-5.30 ± 10.83
Types of DSM	-	-	-
Dome, n (%)	+3 (5%)	-	-
Vertical, n (%)	-3 (5%)	-	-
Horizontal, n (%)	0 (0%)	-	-
BCVA, logMAR ± SD	-0.02 ± 0.02	.56	-0.03 ± 0.14

AL = axial length; BCVA = best-corrected visual acuity; CTh = choroidal thickness; DSM = dome-shaped macula; SD = standard deviation.

<sup>a</sup>P values for the comparison between initial and final measurements using Student *t* test. (Asterisk indicates statistically significant values.)

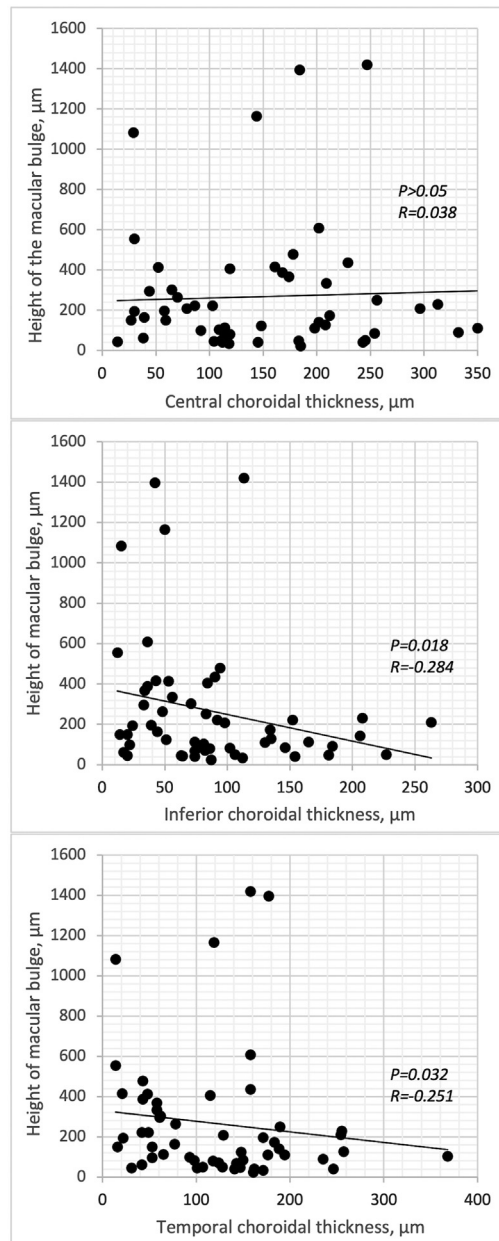


**FIGURE 3.** Anatomic changes during follow-up suggesting that dome-shaped macula (DSM) evolves similarly to other types of myopic staphyloma: axial length and macular bulge increase over time, more severely in more myopic eyes and in eyes with bigger DSM, respectively. Eye elongation correlated with baseline axial length (Top) and increase in macular bulge correlated with baseline macular bulge height (Bottom).

other 32 of 16 patients (55%) were included into the classic DSM group.

The baseline characteristics of the entire population are presented in Table 1. Most of the studied eyes were myopic, with AL superior to 24.00 mm (48 eyes, 82.8%); 10 (17.2%) were emmetropic or hyperopic with a mean RE of  $+1.50 \pm 0.93$  diopters (D) (from 0.00 to +3.00 D). Mean AL in those 10 eyes was  $23.54 \pm 1.17$  mm.

Initially, 9 eyes were pseudophakic and 49 were phakic. A total of 33 were operated on for cataract by phacoemulsification during the follow-up period. None of the eyes underwent vitrectomy. Eight eyes from the classic DSM group had parafoveal retinoschisis and 1 eye from the mini-DSM group showed lamellar macula hole. Those conditions remained stable during follow-up. SRD was observed in 11 eyes (1 eye with mini-DSM and 10 eyes with classic DSM). Fluid spontaneously decreased in 4 eyes (including the one with mini-DSM) and completely resolved in 5. No evolution was observed in the 2 remaining eyes. No CNV



**FIGURE 4.** The height of dome-shaped macula negatively correlated with choroidal thickness (CTh) only in the perimacular areas, suggesting progressive choroidal atrophy and deepening of the peri-dome region. There was no correlation between bulge height and central CTh (Top), while peripheral CTh (inferior and temporal) correlated negatively with macular bulge height (Middle and Bottom).

was associated with SRD and no treatment (ie, intravitreal injection or photodynamic therapy) was conducted.

The mean follow-up period was  $51.76 \pm 36.01$  months. In total, 20 eyes (34.5%) had been followed up for more than 5 years and 5 (8.6%) for more than 8 years. Mean BCVA did not change during the follow-up period. The

**TABLE 3.** Comparisons of Visual and Anatomic Baseline Characteristics, as Well as of Their Evolutions, Between the Mini-dome-shaped Macula and Classic Dome-shaped Macula Group

	Mini-DSM Group	Classic DSM Group	P
Eyes, n (%)	26 (45%)	32 (55%)	-
Subjects, M/F, n (%)	6/11 (35%/65%)	9/10 (47%/53%)	.60
Age, years ± SD	65.92 ± 11.51	63.19 ± 13.78	.422
Bilateral/unilateral condition, eyes (%)	22 (84.6%)/4 (15.4%)	30 (93.8%)/2 (6.3%)	.256
Follow-up period, months ± SD	51.73 ± 27.42	51.78 ± 42.18	.996
RE, diopters ± SD	-5.935 ± 7.505	-9.278 ± 6.668	.05*
AL, mm ± SD	26.17 ± 3.072	27.66 ± 3.07	.027*
Height of the bulge, μm ± SD	60.52 ± 22.69	377.29 ± 315.51	<.001*
Width of the bulge, μm ± SD	3887 ± 1806	6755 ± 1309	<.001*
Type of DSM	-	-	-
Dome, n (%)	15 (57.7%)	15 (46.9%)	.85
Vertical, n (%)	5 (19.2%)	12 (37.5%)	.09
Horizontal, n (%)	6 (23.1%)	5 (15.6%)	.52
Eye elongation per year, mm ± SD	0.003 ± 0.069	0.062 ± 0.17	.007*
Increase of bulge height per year, μm ± SD	4.76 ± 8.85	9.59 ± 15.68	.07
Decrease of central CTh per year, μm ± SD	2.19 ± 7.25	-0.11 ± 7.35	.15
Decrease of nasal CTh per year, μm ± SD	-28.89 ± 25.14	-19.14 ± 26.53	.17
Decrease of temporal CTh per year, μm ± SD	2.063 ± 20.69	-4.83 ± 7.86	.037*
Decrease of superior CTh per year, μm ± SD	-0.36 ± 11.87	-1.31 ± 7.60	.68
Decrease of inferior CTh per year, μm ± SD	-5.32 ± 10.43	-5.28 ± 11.34	.07
BCVA at baseline, logMAR ± SD (Snellen)	0.169 ± 0.172 (20/32)	0.437 ± 0.393 (20/50)	.002*
BCVA change during follow-up, logMAR ± SD	0.026 ± 0.19	-0.060 ± 0.32	.039*
BCVA change per year, logMAR ± SD	0.003 ± 0.059	-0.052 ± 0.17	.044*
Complications	-	-	-
SRD, n (%)	1 (3.8%)	10 (32.3%)	.007*
Central retinal atrophy, n (%)	3 (11.5%)	12 (38.7%)	.02*

AL = axial length; BCVA = best-corrected visual acuity; CTh = choroidal thickness; DSM = dome-shaped macula; F = female; M = male; RE = refractive error; SD = standard deviation; SRD = serous retinal detachment.

Asterisk indicates statistically significant values.

most significant changes for all 58 eyes during follow-up are presented in [Table 2](#).

- **AXIAL LENGTH CHANGES:** The mean AL of the 58 eyes significantly increased by  $0.13 \pm 0.15$  mm ( $P = .010$ ) during follow-up ([Table 2](#)), with a mean increase of  $0.04 \pm 0.13$  mm per year. This elongation was not observed in 20 eyes (34.5%). Baseline AL correlated with mean elongation of the eyes per year ( $R = 0.353$ ;  $P = .003$ ) ([Figure 3](#)) and with mean increase in bulge height per year ( $P = .03$ ). AL correlated negatively with central CTh ( $P < .001$ ). In eyes showing with an elongation during follow-up (66.5%), significantly higher bulge increase was observed compared to those with steady AL ( $40.94 \pm 83.00$  μm vs  $15.42 \pm 24.48$  μm;  $P = .04$ ). Although eyes with steady AL were found in both the classic MB group and mini-MB group, 70% were emmetropic or hyperopic and RE was smaller than in eyes presenting with elongation ( $-4.88 \pm 7.04$  D vs  $-9.5 \pm 6.8$  D;  $P = .026$ ).

- **MACULAR BULGE EVOLUTION:** Mean height of the macular bulge significantly increased over time (from  $235.88 \pm$

$282.44$  μm to  $262.34 \pm 317.15$  μm;  $P < .001$ ) ([Table 2](#)), with a mean increase of  $7.39 \pm 13.14$  μm per year. This increase was not observed in 19 eyes (32%). The initial bulge height correlated positively with mean increase in bulge height per year ( $R = 0.305$ ;  $P = .012$ ) ([Figure 3](#)) and with mean elongation of the eye per year ( $P = .043$ ). Macular bulge height did not correlate with central CTh ( $P > .05$ ), while a negative correlation was observed with inferior and temporal CTh ( $R = 284$ ,  $P = .018$  and  $R = 251$ ,  $P = .032$ , respectively) ([Figure 4](#)). In the 39 eyes (68%) showing bulge increase, this increase correlated with eye elongation during follow-up ( $R = 0.334$ ;  $P = .025$ ). Also, mean bulge increase per year in those eyes correlated with mean eye elongation per year ( $R = 0.332$ ;  $P = .026$ ). Bulge width did not change significantly during the follow-up period. Type of DSM (vertical, horizontal, or round) did not change during the study, except in 3 eyes (5.2%), which changed from vertical to round type ([Table 2](#)).

- **DIFFERENTIAL ELONGATION:** Mean AL change was higher than mean DSM height change with time (i.e.,

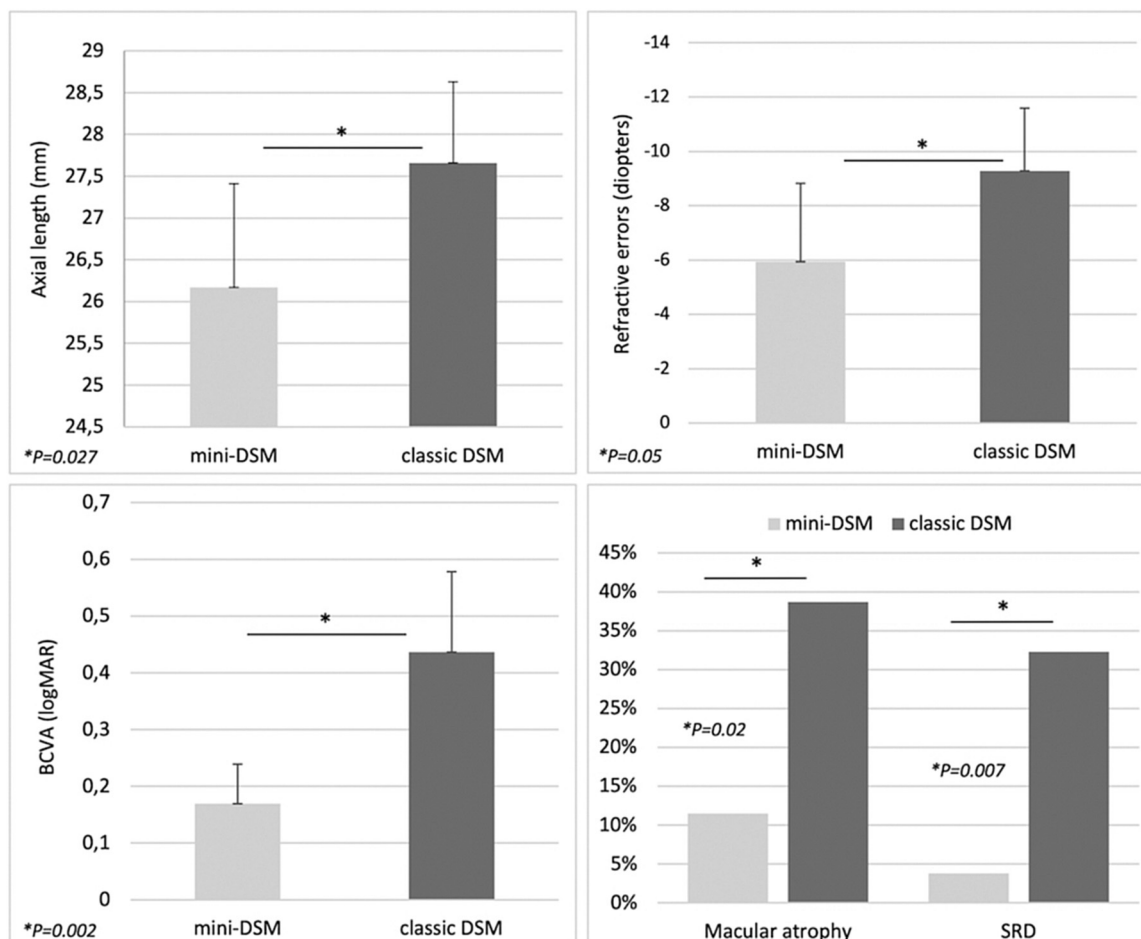


FIGURE 5. Mini-dome-shaped macula (DSM) are associated with less myopic eyes and better visual outcomes than classic DSM. Axial length was reduced in mini-DSM (Top left), as was refractive error (RE) (Top right). Best-corrected visual acuity (BCVA) was greater (Bottom left) and eyes with mini-DSM had less macular atrophy and serous retinal detachment (Bottom right). Asterisk indicate statistically significant differences, with corresponding *P* values indicated in each graph.

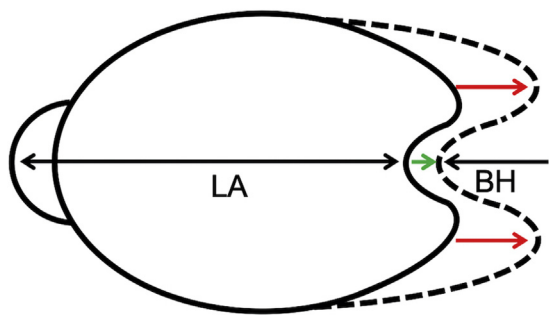
0.13 +/- 0.15 mm vs. 26.46 +/- 34.68  $\mu$ m;  $P < 0.001$ ). Also, mean AL change per year was higher than mean DSM height change per year (i.e., 36 +/- 132  $\mu$ m vs. 7.39 +/- 13.14  $\mu$ m;  $P < 0.001$ ). Mean AL change was compared to peri-dome elongation (i.e., AL change at the top of the bulge + bulge height change). Mean AL change was significantly smaller than peri-dome elongation (i.e., 129 +/- 150  $\mu$ m vs. 158 +/- 377  $\mu$ m;  $P < 0.001$ ). Mean AL change per year was significantly smaller than peri-dome elongation per year (i.e., 36 +/- 132  $\mu$ m vs. 42 +/- 135  $\mu$ m;  $P < 0.001$ ).

• **CHOROIDAL THICKNESS ASSESSMENTS:** Mean central CTh at baseline was significantly higher than mean peripheral CTh: nasal, superior, temporal, and inferior CTh were all significantly thinner than central CTh ( $P < .001$ ,  $P = .005$ ,  $P = .03$ , and  $P < .001$ , respectively) (Table 1). Thicker choroid could still be observed in 1 or 2 peripheral quadrants in 20 eyes (34.5%), as compared to central CTh.

Mean nasal, temporal, and inferior CTh significantly decreased during the follow-up period ( $P = .008$ ,  $P = .026$ , and  $P < .001$ , respectively), while central CTh did not significantly change (Table 2). More specifically, nasal and inferior CTh significantly decreased in round DSM (from  $76.59 \pm 56.77 \mu$ m to  $57.90 \pm 40.69 \mu$ m;  $P = .011$ ; and from  $116.1 \pm 74.19 \mu$ m to  $100.1 \pm 63.97 \mu$ m;  $P = .012$ , respectively), while inferior CTh significantly decreased in horizontal DSM (from  $96.78 \pm 65.38 \mu$ m to  $90.40 \pm 72.75 \mu$ m;  $P = .007$ ) and temporal CTh significantly decreased in vertical DSM (from  $135.5 \pm 73.15 \mu$ m to  $110 \pm 65.92 \mu$ m;  $P = .012$ ).

• **“MINI” VS “CLASSIC” DOME-SHAPED MACULA GROUPS:** Comparisons between the mini-DSM and classic DSM groups showed that mean age, mean follow-up period, mean bulge width, and type of DSM did not differ between groups (Table 3). The DSM were mainly of round type in both groups (57.7% vs 46.9% of eyes in the mini-DSM and classic DSM groups, respectively).





**FIGURE 6.** Differential elongation in eyes with dome-shaped macula (DSM). DSM evolution with time is represented in dotted lines: axial length (AL) modification with time, represented by the green arrow, is greater than macular bulge height (BH) increase (single black arrow pointing left) but the peri-dome elongation (red arrows) is greater than AL change. This supports the fact that eye elongation is more pronounced around the bulge (red arrows) than at the top of the bulge, and that there is a deepening of the perimacular region.

RE was significantly smaller in the mini-DSM group ( $-5.935 \pm 7.51$  vs  $-9.278 \pm 6.67$ ;  $P = .05$ ). Of the 10 of 58 (17.2%) eyes that were emmetropic or hyperopic, 80% were categorized into the mini-DSM group. AL was significantly smaller in the mini-DSM group ( $P = .027$ ).

Mean AL significantly increased in eyes with classic DSM, from  $27.66 \pm 2.69$  to  $27.83 \pm 2.77$  mm ( $P = .007$ ), with a mean elongation per year of  $0.062 \pm 0.17$  mm. In the mini-DSM group, AL increased from  $26.17 \pm 3.07$  mm to  $26.26 \pm 3.0$  mm ( $P > .05$ ), with a mean elongation of  $0.003 \pm 0.069$  per year. This elongation was significantly smaller than in the classic DSM group ( $P = .046$ ) (Table 3). Mean bulge height significantly increased in the mini-DSM group, from  $60.52 \pm 22.69$  mm to  $76.08 \pm 40.56$  mm ( $P = .005$ ), with a mean increase of  $4.76 \pm 8.85$   $\mu\text{m}$  per year, while bulge height increased in the classic DSM group from  $377.29 \pm 315.51$   $\mu\text{m}$  to  $423.77 \pm 362.32$   $\mu\text{m}$  ( $P = .005$ ), with a mean increase per year of  $9.59 \pm 15.68$   $\mu\text{m}$ . Bulge increase in the classic DSM group was greater than in the mini-DSM group; however, this difference was not quite significant ( $P = .07$ ) (Table 3).

The classic DSM group presented with significantly thinner nasal, temporal, superior, and inferior CTh measurements, compared to central CTh ( $P < .001$ ,  $P = .002$ ,  $P = .05$ , and  $P < .001$ , respectively), which was also the case in the mini-DSM group ( $P < .001$ ,  $P = .018$ ,  $P = .04$ , and  $P = .002$ , respectively). In the classic DSM group, inferior, nasal, and temporal CTh significantly decreased during follow-up, from  $88.54 \pm 65.98$   $\mu\text{m}$  to  $80.17 \pm 62.26$   $\mu\text{m}$  ( $P = .005$ ), from  $63.23 \pm 38.87$   $\mu\text{m}$  to  $61.70 \pm 40.73$   $\mu\text{m}$  ( $P = .019$ ), and from  $131.13 \pm 70.78$   $\mu\text{m}$  to  $110.63 \pm 76.29$   $\mu\text{m}$  ( $P < .001$ ), respectively. In the mini-DSM group, however, only inferior CTh significantly decreased during follow-up, from  $112.84 \pm 64.29$   $\mu\text{m}$  to  $95.80 \pm 55.62$   $\mu\text{m}$

( $P = .008$ ). There was no difference observed between central and peripheral CTh decrease per year between the groups; only temporal CTh was observed as more rapidly decreasing in the classic DSM group compared to the mini-DSM group ( $P = .049$ ) (Table 3).

Finally, BCVA in the mini-DSM group was significantly higher compared to the classic DSM group ( $0.169 \pm 0.172$  logMAR vs  $0.437 \pm 0.393$  logMAR;  $P = .002$ ). BCVA decrease per year was significantly greater in the classic DSM group and eyes from the mini-DSM group presented fewer complications, as reported in Table 3 and Figure 5.

## DISCUSSION

IN THIS SERIES OF 58 EYES PRESENTING WITH DSM, MACULAR bulge height increased during a 5-year follow-up period ( $P < .001$ ). Such an increase has already been reported in 2 different studies: Soudier and associates reported a mean increase of  $25.40$   $\mu\text{m}$  after 2 years' follow-up,<sup>6</sup> while Ellabban and associates observed a  $21.1$   $\mu\text{m}$  change during the same follow-up period.<sup>7</sup> In the study presented herein, the mean increase was  $26.46$   $\mu\text{m}$ . In Ellabban and associates' series, both central and peripheral choroidal and scleral thicknesses decreased over time. This decrease was more pronounced in the parafoveal areas, compared to the foveal area, and predominated superiorly, inferiorly, and temporally.<sup>7</sup> Thus, the authors hypothesized that increasing macular bulge height might be the result of parafoveal progressive and asymmetric scleral and choroidal thinning. Some authors have previously stressed the fact that there is a significant difference between central and peripheral CTh.<sup>8,17</sup> The thinning of the parafoveal choroid may reflect the progression of a posterior perimacular staphyloma in highly myopic eyes, according to Soudier and associates, rather than an inward protrusion of the globe, which leads to DSM bulge.<sup>8</sup> Our results are in accordance with those previously reported, as the mean nasal, superior, temporal, and inferior choroid measurements we recorded were all significantly thinner than the central choroid thickness (Table 1). A significant increase of  $0.13$  mm in AL was noted during the study period (Table 2). Bulge increase correlated with AL elongation during follow up ( $R=0.334$ ;  $P=0.025$ ).

The correlation between eye elongation and bulge increase together with the thinning of the peri-dome choroid supports the fact that eye elongation is more pronounced around the bulge than at the top of it. This may suggest that DSM is not due to an inward progressive bulging of the macular area but rather to a differential elongation of the eye predominant in the peri-dome macular region (Figure 6). Increase in AL is one of the characteristics of highly myopic eyes, and it is not surprising that axial elongation was observed in our small cohort in which mainly myopic eyes were present (82.8%).<sup>18</sup> The fact that axial

elongation is higher than bulge increase might suggest that other scleral dynamics located elsewhere than in the posterior pole may participate to eye elongation in eyes with DSM. Other common features of highly myopic eyes were also found in our series, supporting the hypothesis that DSM might represent a particular type of myopic staphyloma in those eyes.<sup>19</sup> Central CTh, for example, correlated negatively with AL, and the increase in such AL was greater in eyes with longer AL, as reported in myopic eyes without DSM by Saka and associates.<sup>18</sup> Of note, Caillaux and associates also reported a positive correlation between macular bulge height and central CTh in DSM eyes<sup>10</sup>; this observation did not support there being a compression of the subfoveal choroid by the posterior sclera, observed in some patients with DSM.<sup>14</sup> In our series, we did not find such a correlation, yet both temporal and inferior CTh did correlate negatively with bulge height (Figure 4), suggesting that choroidal atrophy was present around the macular region and could correspond to a staphylomatous complication. Moreover, peripheral CTh significantly decreased over time in nasal, temporal, and inferior aspects to the foveola, while central CTh remained unchanged (Table 2).

Interestingly, in the present study, DSM was diagnosed in nonmyopic eyes. Indeed, 10 eyes (17.2%) were emmetropic or hyperopic. The majority of those eyes (70%) had a steady AL, along with a smaller increase of the bulge compared to the rest of the eye. Their mean AL was  $23.54 \pm 1.17$  mm, which is relatively high for nonmyopic eyes. DSM in hyperopic eyes has already been reported by Errera and associates in at least 9 eyes of 5 patients.<sup>5</sup> DSM may be underreported in this particular population, since the bulge may be very small and thus not considered as DSM. Indeed, in the present study, the majority of emmetropic or hyperopic eyes had a mini-DSM (80%). The rationale for the differential elongation observed in those eyes is difficult to explain, as they are not presenting with posterior staphyloma, conversely to myopic eyes. It is possible that posterior changes in nonmyopic eyes with DSM do not share the same physiopathology than in myopic eyes. Other studies have already reported DSM eyes without posterior staphyloma. For instance, Errera and associates reported myopic eyes with DSM, of which only 72% showed with posterior staphyloma,<sup>5</sup> and in Imamura and associates' study the presence of posterior staphyloma was not mandatory for DSM diagnosis.<sup>4</sup>

In this study, we included 5 eyes with DSM smaller than  $50 \mu\text{m}$ . As this limit has been proposed in many studies to consider the presence of DSM,<sup>7,11-15</sup> one may argue that those eyes may have biased the results of the present work. Indeed, to exclude this possibility, we performed the statistical analysis after exclusion of those 5 eyes. The results were unchanged (Appendix; Supplemental Material available at [AJO.com](http://AJO.com)).

In previous studies, very small DSM have been poorly studied, even if DSM smaller than  $100 \mu\text{m}$  are currently

seen in daily practice. Those "mini-DSM" might display specific characteristics. In the series presented herein, 26 of 58 eyes (44.8%) exhibited very small bulges (ie,  $<100 \mu\text{m}$ ) and were categorized into a group named the mini-DSM group. The characteristics of this group were different from those of eyes with larger DSM, which we categorized into a second group, named classic DSM: AL was shorter ( $P = .027$ ); RE was smaller ( $P = .05$ ). Axial elongation and bulge increase evolved more slowly. The mini-DSM eyes also presented with higher BCVA ( $P = .002$ ) and fewer complications, such as macular atrophy or SRD ( $P = .02$  and  $P = .007$ , respectively) (Table 3 and Figure 5).

Recently, a specific type of DSM was reported and named RSM.<sup>11</sup> RSM was described in young patients (ie, under 20 years old) with highly myopic eyes. They presented with small macular bulges ( $124 \pm 124 \mu\text{m}$  [range:  $50\text{-}466 \mu\text{m}$ ]) and, compared to "classic DSM," exhibited shorter axial length, higher visual acuity, and fewer myopic maculopathies. In another cohort of myopic children aged under 19, those presenting with DSM also exhibited small macular bulges (ie,  $146 \pm 42 \mu\text{m}$ ) and no associated complications.<sup>20</sup> On analyzing these 2 series, one could wonder whether small DSM in young adults could represent an early stage of DSM. In our series, our patients with mini-DSM shared some characteristics with those reported in the 2 studies cited above (ie, smaller AL), smaller RE, higher visual acuity, and fewer complications than "classic" DSM. However, our series included mini-DSM patients of adult age, not younger than those of the classic DSM group (Table 3). It is thus difficult to conclude whether or not small DSM arises in younger patients, since we only studied an adult population. However, our results demonstrated that mini-DSM evolved very slowly and can be present in an adult population.

Several forms of DSM have been reported in the literature, including horizontal, vertical, or round DSM; RSM; and DSM associated with MBMDs. In this series, we added another type of DSM, named "mini-DSM," defined as exhibiting a macular bulge smaller than  $100 \mu\text{m}$ . These different types of DSM share common features, such as the progressive bulging aspect on OCT and the fact that they are mainly observed in myopic or highly myopic eyes. They also exhibit differences, however, in terms of disease progression and visual prognosis, and their pathogenesis actually might not be exactly the same. In extremely myopic eyes, MBMDs seem to be found quite often and may explain an inward bulging of the sclera. In more moderately myopic eyes, on the other hand, overall asymmetrical thinning of the choroid and sclera resulting from eye lengthening and perimacular staphyloma deepening might lead to bulge increase. In fact, in the series reporting MBMDs, the eyes studied were very elongated (mean AL  $>30$  mm).<sup>9,14</sup> In those cases, thin and fragile sclera may show some specific alterations that provoke choroidal compression, thus leading to increased inward

bulge. Conversely, in both our series and that of Ellabban and associates, mean AL was inferior to 30 mm and no MBMDs were noted.<sup>7</sup>

The retrospective design of this study and limited number of patients represent some limitations. AL measurements were realized using 3 different devices; although all patients underwent their first AL measurement on a Zeiss IOLMaster 500, subsequently, owing to device replacements, the second measurements were performed on either a Zeiss IOLMaster 700 or Tomey OA-2000, depending on the center (Paris or Strasbourg). Good comparability of AL measurement between Zeiss IOLMaster 500 and 700 has been demonstrated, however, by Akman and associates; the mean AL measurement difference between the 2 devices for 1 patient, for example, was  $0.005 \pm 0.02$  mm.<sup>21</sup> Also, high accuracy and repeatability of AL assessments using the Tomey OA-2000 have been reported by Wang and associates, who reported a standard deviation of 0.02 mm for AL measurement in their study.<sup>22</sup> Moreover, the difference in AL reported during follow-up in our series (ie, 0.13 mm) was much higher than those reported risks of errors. Thus, we can be confident there was sufficient comparability in our AL assessments. Another bias could be the bulge height measurement; this was assessed using the manual calipers of the Spectralis OCT and, despite the “follow-up” setting used to optimize measurement repeatability, unavoidable errors may exist owing to the limit of accuracy of the eye tracking system.

In conclusion, macular bulge increase in DSM is associated with an elongation of the affected eye. This finding, along with a global thinning of the perimacular choroid, indicates that DSM evolution is secondary to a differential elongation of the eye predominant in the peri-dome region, rather than to an inward push of retromacular sclera. The minimal height for DSM bulge has not been established yet. Our study shows that in DSM inferior to 100 micrometers, or “mini-DSM,” there is also an evolution, which could lead to greater DSM bulge, as classically described. However, mini-DSM seem to be characterized by better visual prognosis and a smaller rate of complications.

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## CRediT AUTHORSHIP CONTRIBUTION STATEMENT

**LEA DORMEGNY:** CONCEPTUALIZATION, METHODOLOGY, Formal analysis, Investigation, Data curation, Writing - original draft, Visualization. **Xuanli Liu:** Conceptualization, Methodology. **Elise Philippakis:** Resources, Writing - review & editing. **Ramin Tadayoni:** Validation, Supervision. **Zsolt Bocskei:** Data curation. **Tristan Bourcier:** Validation. **Arnaud Sauer:** Resources, Conceptualization, Visualization, Writing - review & editing. **Alain Gaudric:** Project administration. **Claude Speeg-Schatz:** Conceptualization, Formal analysis, Writing - review & editing, Visualization, Supervision.

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FUNDING/SUPPORT: THIS STUDY DID NOT REQUIRE ANY FINANCIAL SUPPORT. FINANCIAL DISCLOSURES: THE AUTHORS HAVE no financial conflicts to disclose. All authors attest that they meet the current ICMJE criteria for authorship.

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