

# Dome and Ridge-Shaped Macula Angle and its Association with Subretinal Fluid

## Ângulo da Dome e *Ridge-Shaped* Macula e sua Associação com Líquido Subretiniano

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### ABSTRACT

**INTRODUCTION:** Dome and ridge-shaped macula (DSM and RSM) are defined by a bulge of retinal pigment epithelium (RPE), choroid, and sclera of more than 50  $\mu\text{m}$  in height above a line connecting the RPE on both sides, in an optical coherence tomography exam (OCT). RSM is defined by a macular elevation only in one meridian across the fovea, whereas the DSM has an elevation in both horizontal and vertical meridians. Their pathogenesis remains controversial. The purpose of this study is to compare OCT parameters in eyes with DSM and RSM with and without subretinal fluid (SRF).

**METHODS:** A retrospective, single-center study was conducted in Centro Hospitalar Universitário de São João (Porto, Portugal). Data collected included age at diagnosis, the affected eye, type of bulge (DSM or RSM), best corrected visual acuity (BVCA), spherical equivalent, intraocular pressure (IOP), subfoveal choroidal thickness (SCT), DSM and RSM angle, and complications.

**RESULTS:** Fifty eyes from 27 patients were evaluated. Twenty-three patients were female (85.19%) and the mean patient age was 53.1 years (range 20–90 years). The condition was bilateral in 23 patients (85.19%). Twenty eyes were identified as RSM and 24 eyes as DSM. In the remaining 6 eyes only, horizontal scans were performed. The mean spherical equivalent was not statistically different between the group without SRF and the group with SRF ( $-11.8 \pm 6.7 \text{ D}$  vs  $-7.5 \pm 6.4 \text{ D}$ ,  $p=0.059$ ). No significant difference was observed in the IOP between both groups ( $p=0.458$ ). The mean SCT was higher in eyes with SRF ( $129.44 \pm 125.96 \mu\text{m}$  vs  $233.63 \pm 99.18 \mu\text{m}$ ,  $p=0.006$ ). DSM/RSM angle was positively correlated to the SRF area ( $p=0.026$ ) but no significant correlation was found between SCT and the SRF area ( $p=0.759$ ). The proportion of eyes with SRF was not different between eyes with DSM and RSM ( $p=0.601$ ). The following complications were found: SRF (N=16), epiretinal membrane (N=9), macular neovascularization (N=8), retinoschisis (N=4), and macular pseudohole (N=3).

**CONCLUSION:** Eyes with DSM/RSM complicated with SRF have a greater SCT. DSM/RSM angle may be a useful OCT marker when evaluating patients with DMS/RSM as it seems to be associated with more SRF.

**KEYWORDS:** Choroid; Macula Lutea; Myopia; Subretinal Fluid; Tomography, Optical Coherence; Visual Acuity.

## RESUMO

**INTRODUÇÃO:** A mácula em cúpula (DSM) e em crista (RSM) são definidas por uma elevação do epitélio pigmentar da retina (EPR), coróide e esclera superior a 50 µm relativamente a um plano que une o EPR no início da elevação, num exame de tomografia de coerência óptica (OCT). Na RSM a elevação macular existe apenas num meridiano, ao contrário da DSM em que a elevação está presente nos meridianos horizontais e verticais. A patogénese destas entidades permanece controversa. O objetivo deste estudo é comparar parâmetros de OCT em olhos com DSM e RSM com e sem líquido subretiniano (LSR).

**MÉTODOS:** Foi realizado um estudo retrospectivo no Centro Hospitalar Universitário de São João (Porto, Portugal). Os dados incluídos no estudo foram idade ao diagnóstico, olho afetado, tipo de DSM/RSM, melhor acuidade visual corrigida (BVCA), equivalente esférico, pressão intraocular (PIO), espessura da coróide subfoveal (ECF), ângulo da DSM/RSM e complicações.

**RESULTADOS:** Foram avaliados 50 olhos de 27 doentes. Vinte e três doentes eram do sexo feminino (85,19%) e a idade média foi de 53,1 anos (20-90 anos). DSM/RSM era bilateral em 23 doentes (85,19%). Vinte olhos foram identificados como RSM e 24 como DSM. Nos restantes 6 olhos foram realizados apenas OCT com corte horizontal. O equivalente esférico médio não foi estatisticamente diferente entre o grupo sem e com LSR (-11,8±6,7 D vs -7,5±6,4 D,  $p=0,059$ ). Não foi observada diferença significativa na PIO entre os dois grupos ( $p=0,458$ ). A ECF média foi maior nos olhos com LSR (129,44±125,96 µm vs 233,63±99,18 µm,  $p=0,006$ ). O ângulo DSM/RSM correlacionou-se positivamente com a área de LSR ( $p=0,026$ ), mas não se identificou correlação entre a ECF e a área de LSR ( $p=0,759$ ). A presença de LSR não foi diferente entre olhos com DSM e RSM ( $p=0,601$ ). Foram encontradas as seguintes complicações: LSR (N=16), membrana epirretiniana (N=9), neovascularização macular (N=8), retinosquias (N=4) e pseudoburaco macular (N=3).

**CONCLUSÃO:** A ECF é superior em olhos com DSM/RSM que apresentam LSR. O ângulo da DSM/RSM pode ser um marcador útil na avaliação de doentes com DSM/RSM, pois parece estar associado a mais LSR.

**PALAVRAS-CHAVE:** Acuidade Visual; Coroide; Fluido Subretiniano; Macula Lutea; Mioopia; Tomografia de Coerência Ótica.

## INTRODUCTION

Dome-shaped macula (DSM) was first described in 2008 and it was characterized by an inward bulge of the macula within the chorioretinal posterior concavity of the eye. The threshold used to define DSM on optical coherence tomography exam (OCT) was a macular bulge height of more than 50 µm above the line tangent to the retinal pigment epithelium (RPE).<sup>1</sup>

Ridge-shaped macula (RSM) has been the term used to define the macular elevation only in one meridian across the fovea, whereas the macular curvature was unremarkable in the perpendicular meridian, different from the typical bidirectional dome (DSM).<sup>2</sup>

Various theories have been discussed about its etiology: hypotonia, tangential vitreomacular traction, resistance of the sclera to the staphylomatous deformation, a localized choroid thickening, a localized scleral thickening, or an adaptive mechanism to minimize defocus in highly myopic eyes undergoing axial length increases.<sup>1,3,4</sup>

DSM was initially assumed to be specific to highly myopic eyes, but a recent report has described that this condition

can also be found in mild myopia or even in hypermetropia.<sup>5</sup>

The main complication of DSM is the occurrence of subretinal fluid (SRF). Its pathogenesis remains unknown, but it is thought to be secondary to increased scleral thickness in DSM eyes, which affects the flow of choroidal fluid. Different therapies have been proposed, including photodynamic therapy, mineralocorticoid-receptor antagonists, aflibercept, and topical carbonic anhydrase inhibitors. However, all studies were case reports or small series and no comparison with a control group was performed. To date, no effective treatment is available for SRF complicating DSM/RSM.<sup>6</sup>

The purpose of this study is to compare OCT parameters in eyes with DSM and RSM with and without SRF.

## METHODS

This retrospective observational study reviewed medical records of patients with DSM/RSM from Centro Hospitalar Universitário São João, in Porto, Portugal. This study adhered to the principles of the Declaration of Helsinki.

## PATIENTS SELECTION

DSM was defined as an inward bulge of the macular RPE  $>50\ \mu\text{m}$  in both the horizontal and vertical sections of the OCT image. RSM was defined as an inward bulge of the macular RPE  $>50\ \mu\text{m}$  in the horizontal or vertical section of the OCT image.

Exclusion criteria included significant media opacity preventing high-quality imaging, previous vitreoretinal surgeries or SRF associated with the presence of macular choroidal neovascularisation (CNV) on optical coherence tomography (OCTA) or fluorescein angiography.

Data collected included age at diagnosis, affected eye, type of bulge (DSM or RSM), best corrected visual acuity (BVCA), spherical equivalent, intraocular pressure (IOP), subfoveal choroidal thickness (SCT), DSM and RSM angle, and presence of complications. This work described the treatment of SRF related to DSM/RSM. Treatment of SRF due to other etiologies, namely CNV, was not within the scope of this work and therefore was not described.

BCVA was assessed using Snellen visual acuity charts or the Early Treatment of Diabetic Retinopathy Study (ETDRS) scale. Visual acuities were converted to logarithm of the minimum angle of resolution (logMAR) equivalents.

The study population was composed of phakic and pseudophakic patients. If the patient has undergone cataract surgery, the spherical equivalent considered was that of the preoperative period. Eyes were classified as low myopes when the spherical equivalent refractive error was  $\leq -0.50\ \text{D}$  and  $> -6.00\ \text{D}$  and high myopes when the spherical equivalent refractive error was  $\leq -6.00\ \text{D}$ .<sup>7</sup>

## OPTICAL COHERENCE TOMOGRAPHY

OCT examination was performed using the Spectralis® (Heidelberg Engineering, Heidelberg, Germany). The SCT was measured with a vertical line perpendicular to the RPE in the foveal region between the hyperreflective RPE layer and the choroid–sclera interface. The presence of SRF was assessed on OCT scans and its area was measured using the Heidelberg tool (Fig. 1). DSM/RSM angle was measured between a line tangent to the outer border of the RPE and a line tangent to the RPE present in the bulge, using the freely available ImageJ software 0.5.7 version (Figs. 2 and 3).

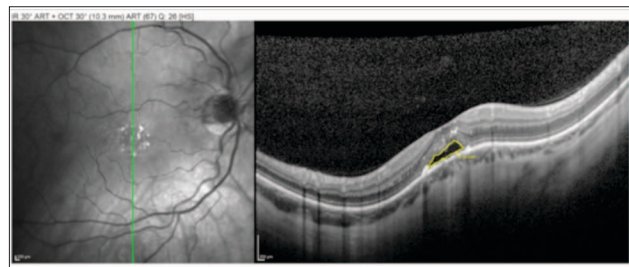


Figure 1. A vertical OS OCT scan showing a DSM with SRF. The SRF area was  $0.06\ \text{mm}^2$ .

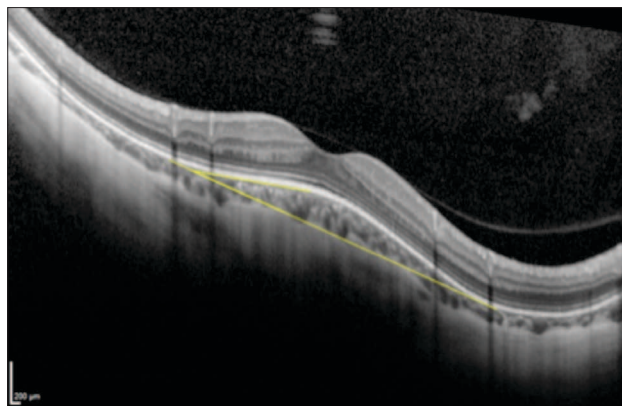


Figure 2. A vertical OS OCT scan showing a DSM without SRF. The DSM angle measured was  $14.57^\circ$ .

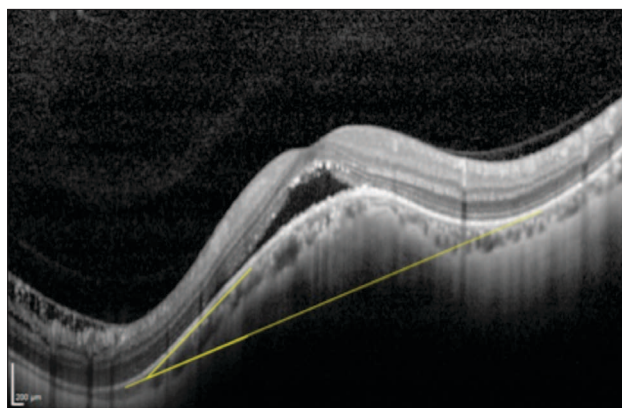


Figure 3. A vertical OD OCT scan showing a RSM with SRF. The RSM angle measured was  $23.85^\circ$ .

## STATISTICAL ANALYSIS

Statistical analyses were performed with SPSS for Windows software (version 28.0, SPSS, Inc., Chicago, IL). A  $p$ -value  $< 0.05$  was considered statistically significant. Normality was assessed using the Shapiro-Wilk test. Continuous variables were compared with parametric or non-parametric tests, according to the normality of data. Chi-square test or Fisher's exact tests were performed for categorical variables.

## RESULTS

Fifty eyes with DSM/RSM from 27 patients were evaluated. The mean patient age was  $53.1 \pm 17.0$  years (range, 20–90 years). Of the 50 eyes, 46 (92.0%) were from female patients, and 26 (52.0%) were right eyes. The condition was bilateral in 23 of 27 (85.2%) patients. The median spherical equivalent was  $-9.06\ \text{D}$  (IQR, 12.66; range,  $-20.75\ \text{D}$  to  $+0.63\ \text{D}$ ). In 12 eyes (24.0%), refraction data were missing from the medical record. The mean SCT was  $162.80\ \mu\text{m}$  (SD, 16.00–487.00  $\mu\text{m}$ ). The mean DSM/RSM angle was  $17.74^\circ$  (SD, 3.34–50.22 $^\circ$ ). Thirty-one eyes (62.0%) were highly myopic eyes, 11 (22.0%) were low myopic eyes, 3 (6.0%) were

emmetropes and 1 (2.0%) was hyperope.

The mean IOP was 17.1±4.0 mmHg, ranging between 12 to 30 mmHg. One patient, with bilateral DSM, had ocular hypertension (OD 30 mmHg, OS 28 mmHg), and hypotensive drops were then started.

Regarding the type of bulge anatomy, 24 eyes were classified as DSM and 20 eyes as RSM on SD-OCT. In the remaining 6 eyes only, horizontal scans were performed. These six eyes were only included in the initial descriptive analysis but were not classified as DSM or RSM and, therefore, were not included in the more specific analysis.

SRF was observed in 16 eyes (32.0%). The mean SRF area was 0.125 mm<sup>2</sup>, ranging from 0.02 to 0.35 mm<sup>2</sup>. Comparing the two groups (with and without SRF), at baseline, no significant differences were found regarding sex ( $p=0.643$ ), age at diagnosis ( $p=0.782$ ), eye laterality ( $p=0.104$ ), BCVA ( $p=0.717$ ) or intraocular pressure ( $p=.458$ ). Although eyes without SRF had a superior absolute value of spherical equivalent, it was not statistically different between the two groups ( $-11.8±6.7$  D *vs*  $-7.5±6.4$  D,  $p=0.059$ ).

The mean SCT was higher in eyes with SRF (129.44±125.96 μm *vs* 233.63±99.18 μm,  $p=0.006$ ), however, no significant correlation was found between SCT and the SRF area ( $p=0.759$ ). DSM/RSM angle was not different between the two groups ( $p=.067$ ), but it positively correlated to the SRF area ( $p=0.026$ ). No significant correlation was observed between the DSM/RSM angle and SCT ( $p=.0862$ ).

The proportion of eyes with SRF was not different between eyes with DSM and RSM ( $p=0.601$ ).

Fisher's exact test was used to determine if there was a significant association between SRF and refractive error (highly myopic eyes, low myopic eyes, and nonmyopic eyes). This analysis demonstrated that there was a statistically significant association between SRF and refractive error ( $p<0.001$ ). In fact, eyes with low myopia were 12.89 times more likely to have SRF than those with high myopia (OR 12.89, 95% CI 2.62-63.31).

We included the significant parameters in the generalized estimating equation, to account for possible inter-eye correlation from the same individual. Regarding the SCT and the refractive error, only the former was found to be a significant factor affecting the presence of SRF ( $p=0.044$ ). DSM/RSM angle continued to be a significant factor affecting the SRF area ( $p<0.001$ ).

The following complications were found: SRF (N=16; 32.0%), epiretinal membrane (N=9; 18.0%), macular neovascularization (N=8; 16.0%), retinoschisis (N=4; 8.0%), and macular pseudohole (N=3; 6.0%). The spherical equivalent was not higher in patients with complications ( $p=0.779$ ).

Of the 16 eyes with SRF, 15 were treated: PDT (3 eyes), anti-VEGF injection (1 eye), PDT + anti-VEGF injection (4 eyes), PDT + oral aldosterone antagonist (eplerenone) (5 eyes), PDT+ anti-VEGF + oral aldosterone antagonist (eplerenone) (2 eyes). In relation to the amount of SRF, it partially/totally improved only in 3 of the 15 treated eyes (one treated only with TFD, one treated with PDT + anti-VEGF injection, and one treated with PDT + oral aldosterone antagonist).

**Table 1. Demographics and baseline characteristics of the study eyes.**

N	50
Patient Sex	46F/4M
Age	53.1±17.0 y (20–90 y)
SE	-9.06 D (IQR, 12.66; -20.75 to +0.63 D)
Refraction	31HM/11LM/3E/1H
IOP	17.1±4.0 mmHg (12-30 mmHg)
SCT	162.80±126.91 μm (16.00-487.00 μm)
DSM	24 (48.0%)
RSM	20 (40.0%)
SRF	16 (32.0%)
SRF area	0.125 mm <sup>2</sup> (0.02-0.35 mm <sup>2</sup> )

F- female; M- male; SE- spherical equivalent; HM- high myope; LM – low myope; E – emmetrope; H- hyperope; IOP- intraocular pressure; SCT- subfoveal choroidal thickness; DSM- dome-shaped macula; RSM – ridge-shaped macula; SRF- subretinal fluid

**Table 2. Demographics, baseline characteristics, and OCT parameters comparison between eyes with and without SRF.**

	Without SRF	With SRF	p-value
N	34 (68.0%)	16 (32.0%)	
Sex	28F/6M	14F/2M	1.000 <sup>a</sup>
Age	52.7±16.6 y (20-79 y)	54.1±18.3 y (28-90 y)	0.782 <sup>b</sup>
Eye laterality	15OD/19OS	11OD/5OS	0.104 <sup>c</sup>
BCVA	0.42±0.38 (0-1.6)	0.46±0.28 (0-1.0)	0.717 <sup>b</sup>
IOP	16.8±3.2 mmHg (12-23 mmHg)	17.9±5.4 mmHg (12-30 mmHg)	0.458 <sup>b</sup>
SE	-11.8±6.7 D (-19.5-0.6 D)	-7.5±6.4 D (-20.8-0.4 D)	0.059 <sup>b</sup>
SCT	129.44±125.96 (16.00-487.00 μm)	233.63±99.18 (87.00-397.00 μm)	0.006 <sup>b</sup>
DSM/RSM angle	15.97±11.17° (3.34-50.22°)	21.50±5.45° (10.23-29.90°)	0.067 <sup>b</sup>

F- female; M- male; BCVA- best corrected visual acuity; IOP- intraocular pressure; SE- spherical equivalent; SCT- subfoveal choroidal thickness; DSM- dome-shaped macula; RSM- ridge-shaped macula; a- Fisher exact test; b- T-student test; c- Chi-square test

## DISCUSSION

DSM was first described, in 2008, by Gaucher *et al* in eyes with myopic posterior staphyloma. More recently, in 2013, DSM was differentiated from RSM by Caillaux *et al*. Using horizontal and vertical SD-OCT scans and 3D macular map reconstruction, they described three morphologic DSM patterns: type one - round domes; type two - horizontally oriented oval-shaped domes; type three - vertically oriented oval-shaped domes. Types 2 and 3 were defined as RSM.<sup>1,2,8</sup>

We identified 24 eyes with DSM and 20 with RSM, almost a 1:1 proportion. In a study that evaluated 49 high myopic eyes, the proportion was slightly different: 75.5%

RSM eyes and 24.5% DSM eyes. Because macular bulge in RSM occurs only in one direction, it is important to perform multidirectional SD-OCT scans with at least one horizontal and one vertical scan to diagnose this condition correctly, especially in myopic patients.<sup>9</sup>

Similar to other studies, DSM/RSM was most frequently observed in female patients (N=46; 92%). Gaucher *et al* found a female frequency of 80%, Immamura *et al* 67%, Lorenzo *et al* 66.7%, Garcia-Zamora *et al* 58.3% in DSM and 62.1% in RSM, Negrier *et al* 55.6%.<sup>1,4,9-11</sup>

As found by Errera *et al* in 2014, we also diagnosed this finding in non-myopes eyes – three eyes (6.0%) were emmetropes and one (2.0%) was hyperope. This fact does not fit with the theory that defends that DSM/RSM can be an anatomical adaptation for a better focus of the image on the macula.<sup>5</sup>

Although several studies have been conducted in recent years, the reason why only some of these eyes develop SRF has not yet been understood. We found no differences in terms of sex, age at diagnosis, eye laterality, BCVA, or intraocular pressure between eyes with and without SRF. In agreement with our results, Lorenzo *et al* found no baseline differences between eyes with or without SRF in terms of sex, age, eye laterality, the orientation of DSM, presence of amblyopia, total follow-up time, or initial BCVA.<sup>10</sup>

SRF treatment included intravitreal injections of bevacizumab (1.25 mg/0.05 mL) or aflibercept (2.0 mg/0.05 mL) and/or photodynamic therapy with half of the standard dose (3 mg/m<sup>2</sup>) and/or oral therapy with an aldosterone antagonist. It was decided individually, and no specific guidelines were in place, so we decided not to invest in this analysis.

Regarding the choroid, in our study, mean SCT was higher in eyes with SRF (129.44 ± 125.96 μm *vs* 233.63 ± 99.18 μm, *p*=0.006). A similar conclusion was reached by Errera *et al* in which, eyes with ≥-6 D with SRF or leaks on FA had evidence of increased choroidal thickness compared with those without SRF. However, Negrier *et al* found no significant difference in choroidal thickness in the subfoveal, nasal, temporal, superior, and inferior areas between eyes with and without SRF.<sup>5,11</sup>

We observed that low-myopic eyes were more likely to have SRF than high-myopic eyes. Lorenzo *et al* also demonstrated that the only factor in DSM eyes that was significantly associated with SRF was a lower degree of myopia. Indeed, we know that choroidal thickness tends to decrease with increasing myopia/axial length, as has been described, for example, by Muhiddin HS *et al* in 2022. This difference, taking into account the magnitude of myopia, is in agreement with another of our results – eyes with SRF showed greater SCT. It is possible that the greater the choroidal thickness, the greater the vascular density and therefore the risk of developing SRF. Confirmation of this hypothesis could be carried out by performing optical coherence tomography angiography in these patients. However, to date, existing technology does not provide good-quality scans that effectively quantify choroidal vascular density without the presence of significant artifacts in high-myopic eyes. With the improvement of image acquisition techniques, it may be possible soon.<sup>10,12</sup>

In our study, we reported a novel OCT marker, the DSM/RSM angle. This analysis was performed by transposing a subfoveal OCT section into the ImageJ program. DSM/RSM angle was measured between a line tangent to the outer border of the RPE and a line tangent to the RPE present in the bulge, as can be seen in Figs. 1 and 2. Although DSM/RSM angle was not significantly different between the groups with and without SRF (*p*=0.067), it was positively correlated to the SRF area (*p*=0.026). As the angle measurement could be a prognostic factor in terms of complications or even therapeutic response, its inclusion to OCT software would be a useful and practical tool to improve our results in clinical practice.

As far as we know, our study was the first to measure the SRF area.

Moreover, we observed that patients with a greater SCT were more likely to have SRF and that DSM/RSM angle positively correlated to the area of SRF. Therefore, we must pay particular attention to patients who combine these characteristics. This knowledge can influence our clinical practice, namely the frequency of reevaluation of these patients.

The limitations of the present study include the fact that this was a retrospective study. In addition, other variables that could be associated with complications such as the axial length, the presence/type of staphyloma, and the thickness of the sclera were not evaluated.

## CONCLUSION

Eyes with DSM/RSM complicated with SRF seem to have a greater SCT and lower myopia. DSM/RSM angle may be a useful OCT marker when evaluating patients with DSM/RSM as it seems to be associated with more SRF. Additional research is needed to improve our knowledge regarding this condition and the factors associated with SRF.

## CONTRIBUTORSHIP STATEMENT / DECLARAÇÃO DE CONTRIBUIÇÃO:

JSO: Data collection, literature review, writing, revision.

SP, AC, STC: Writing, revision.

FFR: Revision.

All authors approved the final version to be published.

## RESPONSABILIDADES ÉTICAS

**Conflitos de Interesse:** Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.

**Fontes de Financiamento:** Não existiram fontes externas de financiamento para a realização deste artigo.

**Confidencialidade dos Dados:** Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.

**Proteção de Pessoas e Animais:** Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia revista em 2013 e da Associação Médica Mundial.

**Proveniência e Revisão por Pares:** Não comissionado; revisão externa por pares.

## ETHICAL DISCLOSURES

**Conflicts of Interest:** The authors have no conflicts of interest to declare.

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**Confidentiality of Data:** The authors declare that they have followed the protocols of their work center on the publication of data from patients.

**Protection of Human and Animal Subjects:** The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki as revised in 2013).

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