



# A Child With Dome-Shaped Maculopathy and Spontaneous Resolution Of Subretinal Fluid and Improvement of Vision 6 Years After Initial Diagnosis

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Received: 📅 September 6, 2022

Published: 📅 September 13, 2022

## Abstract

**Purpose:** To report a case of subretinal fluid in dome-shaped maculopathy with spontaneous resolution and consequent improvement of vision up to six years after diagnosis. Additionally, we performed a mini-review of the literature on the treatment of other cases of dome-shaped maculopathy with associated subretinal fluid.

**Methods:** A 9-year-old patient with a history of high myopia complained about metamorphopsia, dyschromatopsia and blurred vision due to dome-shaped maculopathy with subretinal fluid. The right eye received one intravitreal injection of anti-vascular endothelial growth factor, with no effect. The PubMed database was searched between January 2020 and December 2022 for cases reporting on dome-shaped macula associated subretinal fluid and possible treatments.

**Results:** In our case spontaneous resolution of subretinal fluid, with concurrent recovery of visual acuity, occurred after four and six years, respectively in the left and the right eye.

**Conclusion:** The pathogenesis of dome-shaped maculopathy and the development of associated subretinal fluid is still unclear and there is no consensus or high-level evidence on an efficacious treatment. Our case shows that treatment is not always required, and spontaneous resolution can occur, even after several years.

**Summary statement:** Spontaneous resolution of SRF in children with DSM does occur and may be observed after several years, with concurrent improvement in vision to near normal levels. Hence, treatment for SRF in DSM should be chosen with care.

**Keywords:** Case Report; Dome-Shaped Macula; Serous Foveal Detachment; Treatment

**List of Abbreviations:** BCVA: best corrected visual acuity; CMT: central macular thickness; CSR: central serous retinopathy; DSM: dome-shaped maculopathy; OCT: optical coherence tomography; RPE: Retinal pigment epithelium; SRF: subretinal fluid; VEGF: vascular endothelial growth factor

## Introduction

In 2008 Gaucher et al, first reported on a bulge-like protrusion visualized on optical coherence tomography (OCT) imaging and introduced it as dome-shaped macula (DSM) [1]. It was defined as a bulge of retinal pigment epithelium (RPE), choroid, and sclera  $>50\mu\text{m}$  in height above the line connecting the RPE on both sides of the bulge [2]. Because of its predominant occurrence in myopic eyes, the dome is hypothesized to be caused by a local thickening of the sclera that would develop in an effort to minimize the refractive error and achieve better vision. However, cases in mildly myopic, emmetropic or even, although rare, hypermetropic eyes, suggest the involvement of other, possibly genetic, factors. Because the bulge in children with DSM is smaller and has a smoother slope, it is thought that the protrusion is not growing inward, but rather the edges are growing outward as the axial length of the eye increases with age. It is usually an asymptomatic feature [3]. Symptoms mostly occur when dome related complications arise. Possible complications are subretinal fluid (SRF), choroidal neovascularization, macular holes, retinoschisis, epiretinal membrane and vitreomacular traction,

with SRF being the most frequent and the only complication for which DSM has been a proven independent risk factor [4,5]. Since SRF can cause a decrease in visual acuity (VA), knowing how to manage it can be vital. However, there is a severe lack of literature regarding treatment for SRF in DSM, especially in children. We report a case of DSM-related SRF with spontaneous resolution and recovery of VA.

## Case

In 2008 a 5-year-old female child presented to our clinic due to high myopia, with OD -11,25 and OS -9,50 (retinoscopy), and a small esophoria, for which she received glasses and occlusion therapy. Although vision initially developed normal, at the age of 9, she reported seeing strange shapes and colors with best corrected visual acuity (BCVA) of 20/30 Snellen for the right eye and 20/25 Snellen for the left eye. OCT showed DSM with macular SRF in both eyes (Figures 1 & 2). Color fundus photography showed a possible retinal hemorrhage in the right eye (Figure 3). In Figure 4 a fundus photograph of the left eye for reference is shown. Fluorescein angiography (FA) showed signs of possible leakage (Figures 5 & 6).



**Figures 1 & 2:** OCT image of right and left eye respectively showing dome-shaped macula, as defined by Ellabban et al.(1) with subretinal fluid (indicated with an asterisk).

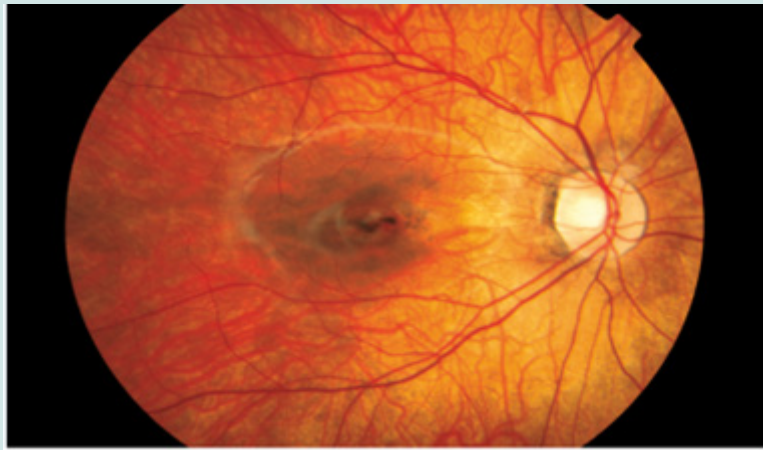


Figure 3: Colour fundus photograph of the right eye, showing possible hemorrhage, patient age 9.

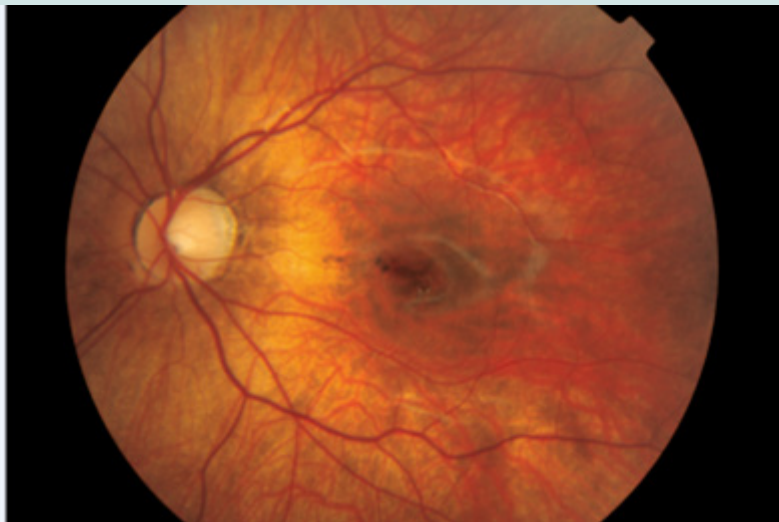


Figure 4: Colour fundus photograph of the left eye for reference, patient age 9.



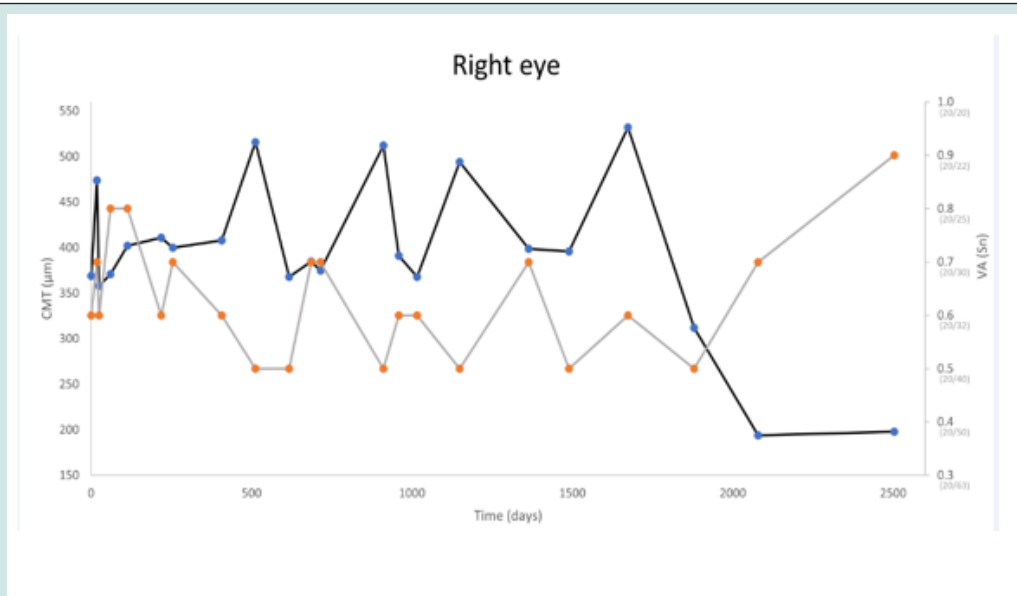
Figure 5: Right eye, early phase fluorescein angiography showing possible leakage, patient age 9.

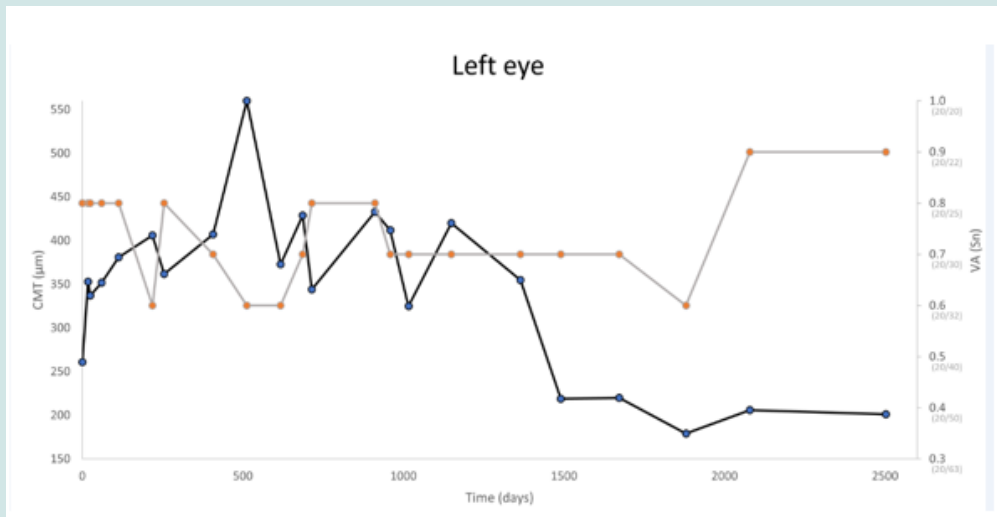


**Figure 6:** Right eye, late phase fluorescein angiography showing possible leakage, patient age 9.

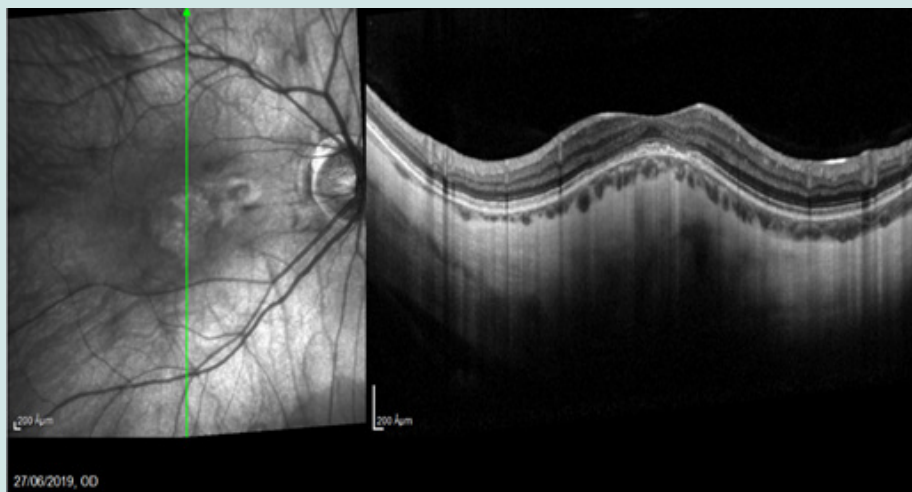
Given these observations a choroidal neovascularization could not be ruled out and the patient consequently received one intravitreal injection of anti-vascular endothelial growth factor (VEGF) in the right eye under general anesthesia. This had no effect on SRF and the diagnosis of neovascularization was discarded thereafter. The following two years the patient received biannual follow-ups, BCVA remained stable at 20/25 Snellen in both eyes, although SRF and central macular thickness (CMT) increased gradually on OCT. By 2015 BCVA declined to 20/40 Snellen for the right eye and 20/32 Snellen for the left eye. Besides the increase

in SRF and CMT, no other causes were found. Thereafter BCVA fluctuated between 20/40 and 20/30 Snellen for the right eye and 20/32 and 20/25 Snellen for the left eye, largely corresponding to fluctuations in CMT (Figures 7 & 8). SRF in the left eye decreased in 2016 and completely resolved by 2017, with the right eye following suit in 2019 (Figures 9 & 10). This resulted in a marked improvement of vision to 20/22 Snellen for both eyes by the end of 2019. Since then, vision has remained stable, with all subsequent OCTs showing DSM with no signs of SRF and stable normal CMT.

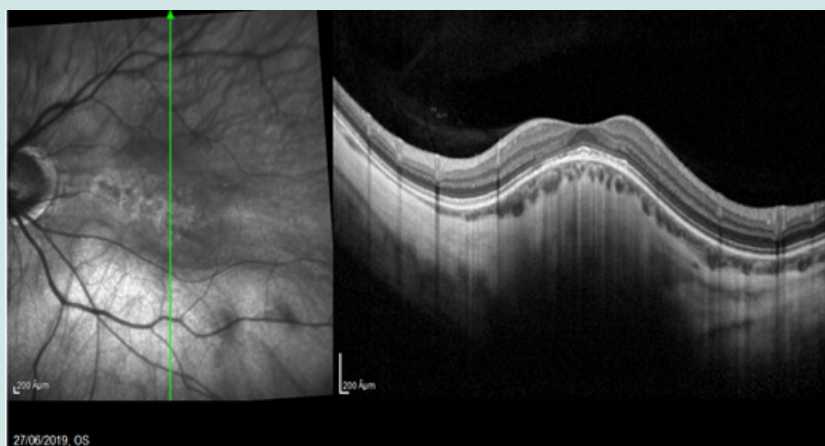




**Figures 7 & 8:** Two graphs depicting the relationship between central macular thickness and visual acuity in the right and left eye respectively. Black lines represent central macular thickness in  $\mu\text{m}$ . Grey lines represent best corrected visual acuity (VA) in decimals (1.0 corresponds to 20/20 Snellen; conversions are approximated to the nearest Snellen equivalent).



**Figures 9:** OCT image of left eye at the age of 13, showing no signs of subretinal fluid.



**Figure 10:** OCT image of right eye at the age of 15, showing no signs of subretinal fluid.

## Discussion

There are several theories regarding the origin of SRF in DSM: 1) increased scleral thickness could possibly obstruct the outflow of choroidal fluid and thus cause the build-up of SRF; 2) increased subfoveal choroidal thickness may result in a CSR-like phenotype; 3) SRF might be secondary to RPE dysfunction; 4) abrupt changes in choroidal thickness and consequently altered choroidal flow could result in SRF; 5) a more recent hypothesis, intervortex venous anastomosis in pachychoroid-related disorders could affect the venous drainage of the retina, which would result in choriocapillaris hypertension and hyperpermeability of the retinal veins, causing SRF. [3,5,6] The association between certain choroidal and dome features and the presence of subretinal fluid has been investigated, with contradicting results. The most recent study by Negrier et al found no association between CT, vessel pattern or dome axis and SRF [7]. All suggested causes of SRF in DSM remain hypothetical and we also lack sufficient evidence regarding the cause of SRF in the here presented case.

Since its pathophysiology is not well understood, treating SRF can be quite difficult. Several treatment options have been tried with limiting success and a varying effect on BCVA. In the table below (Table 1), we summarized cases regarding treatment for DSM with SRF in children between 1990 and 2022. Therapies that have been tried for subretinal fluid in DSM are intravitreal anti-VEGFs, aldosterone antagonists, carbonic anhydrase inhibitors, photodynamic therapy, and laser photocoagulation. These treatments can be expensive or burdensome, hold the risk of complications and have not proven to be effective.

## Conclusion

We here presented a case and showed that spontaneous resorption of SRF in DSM does occur and may be observed after

several years, with concurrent improvement in vision to near normal levels. Hence, treatment for SRF in DSM should be chosen with care.

## Patient Consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

## Conflict of Interest

The authors declare that they have no competing interests.

## References

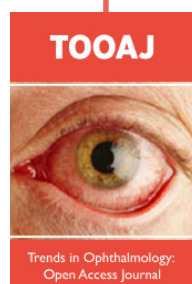
1. Gaucher D, Erginay A, Lecleire-Collet A, Pascale Massin, Alain Gaudric, et al. (2008) Dome-Shaped Macula in Eyes with Myopic Posterior Staphyloma. *Am J of Ophthalmol* 145(5): 909-914.
2. Ellabban AA, Tsujikawa A, Matsumoto A, Masahiro Miyake, Hussein S Elnahas et al. (2013) Three-Dimensional Tomographic Features of Dome-Shaped Macula by Swept-Source Optical Coherence Tomography. *Am J Ophthalmol* 155(2): 320-328.
3. Viola F, Dell'Arti L, Benatti E, Invernizzi A, Giulio Barteselli, et al. (2015) Choroidal Findings in Dome-Shaped Macula in Highly Myopic Eyes: A Longitudinal Study. *Am J Ophthalmol* 159(1): 44-52.
4. Kumar V, Verma S, Azad SV, Abhidnya Surve, Rajpal Vohra, et al. (2020) Dome-shaped Macula- Review of Literature. *Surv Ophthalmol* 66(4): 560-571.
5. Lee GW, Kim JH, Kang SW, Kim J (2020) Structural profile of dome-shaped macula in degenerative myopia and its association with macular disorders. *BMC Ophthalmol* 20(1): 202.
6. Spaide RF, Ledesma-Gil G, Gemmy Cheung CM (2021) Intervortex venous anastomosis in pachychoroid-related disorders. *Retina* 41(5): 997-1004.
7. Negrier P, Couturier A, Gaucher D, Alain Gaudric, Elise Philippakis, et al. (2022) Choroidal thickness and vessel pattern in myopic eyes with dome-shaped macula. *Br J Ophthalmol* 106(12): 1730-1735.



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DOI: [10.32474/TOOAJ.2023.04.000180](https://doi.org/10.32474/TOOAJ.2023.04.000180)



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