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THE IMPACT OF CYSTOID MACULOPATHY IN *USH2A* RETINITIS PIGMENTOSA

A Retrospective 5-year Analysis

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Purpose: Retinitis pigmentosa (RP) is often complicated by cystoid maculopathy (CM). The clinical impact of CM in the RP long-term outcome is still unclear. We described the 5-year outcome of *USH2A*-associated RP eyes with and without CM.

Methods: We retrospectively analyzed patients with *USH2A*-associated RP. Eyes with CM were treatment naïve. We collected data about best-corrected visual acuity, ellipsoid zone (EZ) loss rate, and CM behavior over 5 years of follow-up. The main outcome measures were the impact of CM on best-corrected visual acuity and EZ loss rate. Secondary outcome was the assessment of the clinical impact of CM persistence or regression.

Results: We included 25 RP eyes with CM and 25 eyes without CM. No difference in age and gender distributions have been found. Visual acuity was similar between eyes with and without CM ($P < 0.05$), although it resulted slightly worse in eyes with persistent CM. Eyes without CM had significantly lower EZ width compared with eyes with CM. However, the rate of EZ loss/year was statistically similar between eyes without and with CM. The EZ loss rate maintains similar profiles of progression, up the CM regression, associated with a significant acceleration of EZ loss. After this episode, EZ loss rate showed similar trends than those registered before the CM regression. CM regression and recurrence showed no significant effect on best-corrected visual acuity ($P > 0.05$). Moreover, we found no significant impact of CM reappearance on the EZ loss rate.

Conclusion: CM showed no negative impact on the long-term visual outcome of *USH2A*-associated RP.

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Retinitis pigmentosa (RP) represents the most common inherited retinal dystrophy, characterized by a rod–cone centripetal degeneration. RP eyes are often characterized by the onset of cystoid macular edema with an estimated prevalence of approximatively 10% to 50% of cases.[1,2] RP-related macular edema shows peculiar characteristics, including variable response to topical, systemic and intravitreal treatments, and relative visual acuity stability.[3–6] This is also defined as cystoid maculopathy (CM), since it seems to be not directly caused by blood–retinal barrier breakdown (BRB).[7] Indeed, the pathogenesis is still poorly understood, including a wide spectrum of hypotheses: BRB breakdown, retinal pigment epithelium (RPE) dysfunction, macular Müller cells impairment, autoimmune-related phenomena, and vitreous traction.[3,7]

The clinical impact of CM in RP is still unclear. A recent study showed no relevant effect on the 1-year visual outcome of patients with RP, both considering visual acuity and retinal sensitivity.[8]

The main aim of the present study was to assess the long-term outcome of CM in a cohort of *USH2A*-associated RP.

Materials and Methods

We designed the study as retrospective, observational. The follow-up was 5 years. We recruited

patients affected by *USH2A* RP (Retinal Heredodystrophy Unit, Department of Ophthalmology, IRCCS San Raffaele Scientific Institute, Milan, Italy). The study received the approval by the ethical committee of IRCCS San Raffaele Scientific Institute (MIRD2020). A signed informed consent was obtained from all the patients before the inclusion.

The inclusion criteria were the genetically confirmed diagnosis of *USH2A* RP and age >18 years, with at least one complete ophthalmologic examination for at least 5 consecutive years. The exclusion criteria were macular atrophy, media opacities, other retinal and/or optic nerve diseases (e.g., age-related macular degeneration, uveitis, diabetic retinopathy, glaucoma), and uncontrolled systemic diseases potentially affecting the results of the analyses. We have also carefully inspected the vitreoretinal interface features to exclude those eyes displaying tractional epiretinal membrane (ERM). We have considered only one randomly selected eye for the statistical analysis. All the patients were treatment naïve.

The ophthalmologic assessment included best-corrected visual acuity (BCVA), performed by means of the standard Early Treatment Diabetic Retinopathy Study (ETDRS) chart, anterior and posterior segment slit-lamp evaluation, and applanation tonometry.

Multimodal retinal imaging included blue-light fundus autofluorescence (FAF) and optical coherence tomography (OCT) (Spectralis HRA2+OCT, Heidelberg Engineering; Heidelberg, Germany). OCT acquisition protocol included radial, raster, and dense foveal scans with enhanced depth imaging (EDI).

We extracted the automatic measurement of central macular thickness (CMT) provided by HEYEX2 software. Two expert ophthalmologists (AA, EA) performed the measurement of the ellipsoid zone (EZ) width both on horizontal and vertical foveal OCT scans. Choroidal thickness (CT) was obtained from an horizontal, foveal-centered scan, considering five standardized measures performed under the fovea, at 750 μm distance and at 1,500 μm distance from the fovea, both on the temporal and nasal sides. The mean value was considered for the statistical analyses. We have also included the qualitative assessment of the hyperreflective ganglion cell layer band (HGB).[9]

We calculated the overall EZ band reduction over time and the mean EZ loss rate per year.

A second model has been developed to assess the impact of CM regression on the EZ loss rate. We classified RP eyes with CM in two categories: 1) CM persistence with fluctuations and without CM regression over the follow-up (CM-P) and 2) CM with recurrent behavior and possible complete regression over the follow-up (CM-R). For this subanalysis, we included only eyes showing at least two consecutive annual follow-ups before and after the time point of CM regression. Another mandatory criterion is the presence of only one episode of complete CM regression. This choice was done to standardize the time points for all the included eyes, measuring the changes of EZ width and how CM regression affected on that. On this basis, we analyzed the trend of EZ loss before, during, and after CM regression, comparing the EZ changes in these subgroups of RP eyes.

For statistical purposes (SPSS software package, SPSS, IL), age and gender have been considered as fixed factors. We have checked the normality distribution of each variable through frequency histograms and quantile–quantile plots. Continuous variables have been reported as mean ± SD. Frequency and proportions have been reported as categorical variables. We included only one randomly selected eye. Continuous variables were analyzed by means of a two-tailed *t*-test. Tau–Kendall correlation analysis was used to test the relationships between all the variables considered. One-way ANOVA has been used to test intergroups differences. The Bonferroni correction has been used to address the multiple comparisons. The agreement between the two graders has been assessed by calculating an intraclass correlation coefficient (ICC). The main outcome measure was the EZ loss rate in RP eyes with and without CM. Additional outcome measure was the impact of CM complete regression on EZ loss rate.

Results

We collected data from 25 eyes without CM (25 patients; 13 females; mean age 48 ± 12 years) and 25 eyes with CM (bilateral in 25/25 cases; 100%). These latter were categorized as CM-P (10 patients; five

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females; mean age 51 ± 13 years) and CM-R (15 patients; eight females; mean age 49 ± 8 years). No significant difference in age and gender distributions have been found among the three subgroups ($P > 0.05$). Interestingly, the recurrence of CM in the subgroup with complete regression has been registered in 100% of cases, with a mean recurrence time of 1 ± 1 year since the regression. The overall ICC value for the quantitative measures was 0.90 (range 0.89–0.93). The pathogenic variants are reported in Table 1. All the cases have been classified as nonsyndromic *USH2A* RP. All the patients have never been treated with topical or oral medications for CM during the entire follow-up.

LogMAR BCVA was similar between eyes with and without CM, both considering baseline values (0.1 ± 0.1 LogMAR score vs. 0.0 ± 0.1 LogMAR score (Snellen equivalent 20/25 vs. 20/20); $P > 0.05$) and 5-year follow-up (0.1 ± 0.2 LogMAR score vs. 0.0 ± 0.2 LogMAR score (Snellen equivalent 20/25 vs. 20/20); $P > 0.05$). Eyes without CM started with a significantly lower EZ width at baseline, about eyes with CM, both considering horizontal EZ ($P < 0.001$) and vertical EZ ($P < 0.001$). However, the rate of EZ loss/year was statistically similar between eyes without and with CM (Figure 1), resulting as follows: horizontal EZ (-103 ± 68 μm vs. -142 ± 82 μm; $P > 0.05$) and vertical EZ (-105 ± 82 μm vs. -126 ± 86 μm; $P > 0.05$).

The separated analysis considering RP without CM versus RP with CM-P and RP with CM-R is extensively shown in Table 2.

LogMAR BCVA was similar among the three subgroups. RP eyes with CM-R showed the biggest EZ width, both considering baseline and 5-year follow-up, about the other subgroups ($P < 0.001$). Moreover, RP eyes with CM-P showed significantly bigger EZ width, both considering baseline and 5-year follow-up, about RP eyes without CM ($P < 0.001$).

RP eyes with CM-R were characterized by the highest EZ rate loss (all $P < 0.001$), whereas RP eyes with CM-P showed the lowest EZ rate loss (all $P < 0.001$).

ERM without tractional component was found as follows: RP without CM (32%), RP with CM-P (10%), and RP with CM-R (40%). HGB characterized RP without CM (20%), RP with CM-P (10%), and RP with CM-R (33%).

Fourteen out of 15 RP eyes with complete regression met the inclusion criteria to perform the separated analysis of EZ rate loss among the three RP subgroups. The results are plotted in Figure 2. The EZ loss rate maintains similar profiles of progression, up the CM regression, where it is possible to register a significant acceleration of EZ loss. After this episode, EZ loss rate maintains similar trends than those registered before the CM regression. CM regression and recurrence were not associated with significant BCVA changes ($P > 0.05$). Moreover, we found no significant impact of CM reappearance on the EZ loss rate of the analyzed eyes ($P > 0.05$). If RP with CM

Table 1. Pathogenic Variants of the Recruited Cohort of *USH2A* RP eyes

Patient	Variant 1	Variant 2
1	c.2299del	c.2276G>T
2	c.15089C>A	c.2296T>C
3	c.2276G>T	c.949C>A
4	c.2296T>C	c.2276G>T
5	c.908G>A	c.2296T>C
6	c.908G>A	c.2276G>T
7	c.2276G>T	c.2330G>A
8	c.2276G>T	c.5858C>G
9	c.2276G>T	c.2296T>C
10	c.12559C>T	c.7339A>C
11	c.3332T>G	c.10437G>T
12	c.2296T>C	c.9329C>T
13	c.802G>A	c.12712T>C
14	c.13392G>A	c.9424G>T
15	c.1841-2A>G	c.2299delG
16	c.11235C>A	c.8655_8681+1681del
17	c.2276G>T	c.1841-2A>G
18	c.3938delA	c.10702A>T
19	c.2299del	c.8232G>A
20	c.2299del	c.9424G>T
21	c.653T>A	c.2276G>T
22	c.15020C>T	c.2071T>C
23	c.2276G>T	c.10712C>T
24	c.1036A>G	c.2296T>C
25	c.10712C>T	c.9424G>T
26	c.485+5G>A	c.10817T>C
27	c.5418_5424del	c.13486G>A
28	c.13392G>A	c.2276G>T
29	c.2276G>T	c.13477C>T
30	c.5418_5424del	c.(784+1_785–1)_(1840+1_1841–1)del
31	c.10436G>A	c.5776+1G>A
32	c.2710_2720dup	c.1055C>T
33	c.2276G>T	c.2296T>C
34	c.2296T>C	c.10712C>T
35	c.2276G>T	c.1841-2A>G
36	c.2276G>T	c.9424G>T
37	c.2299del	c.2276G>T
38	c.908G>A	c.2276G>T
39	c.908G>A	c.2296T>C
40	c.802G>A	c.14219C>A
41	c.13392G>A	c.2276G>T
42	c.2299del	c.5418_5424del
43	c.15089C>A	c.2296T>C
44	c.3332T>G	c.908G>A
45	c.2299del	c.2276G>T
46	c.2276G>T	c.2299del
47	c.2710_2720dup	c.908G>A
48	c.2299del	c.5418_5424del
49	c.2276G>T	c.2296T>C
50	c.3332T>G	c.1055C>T

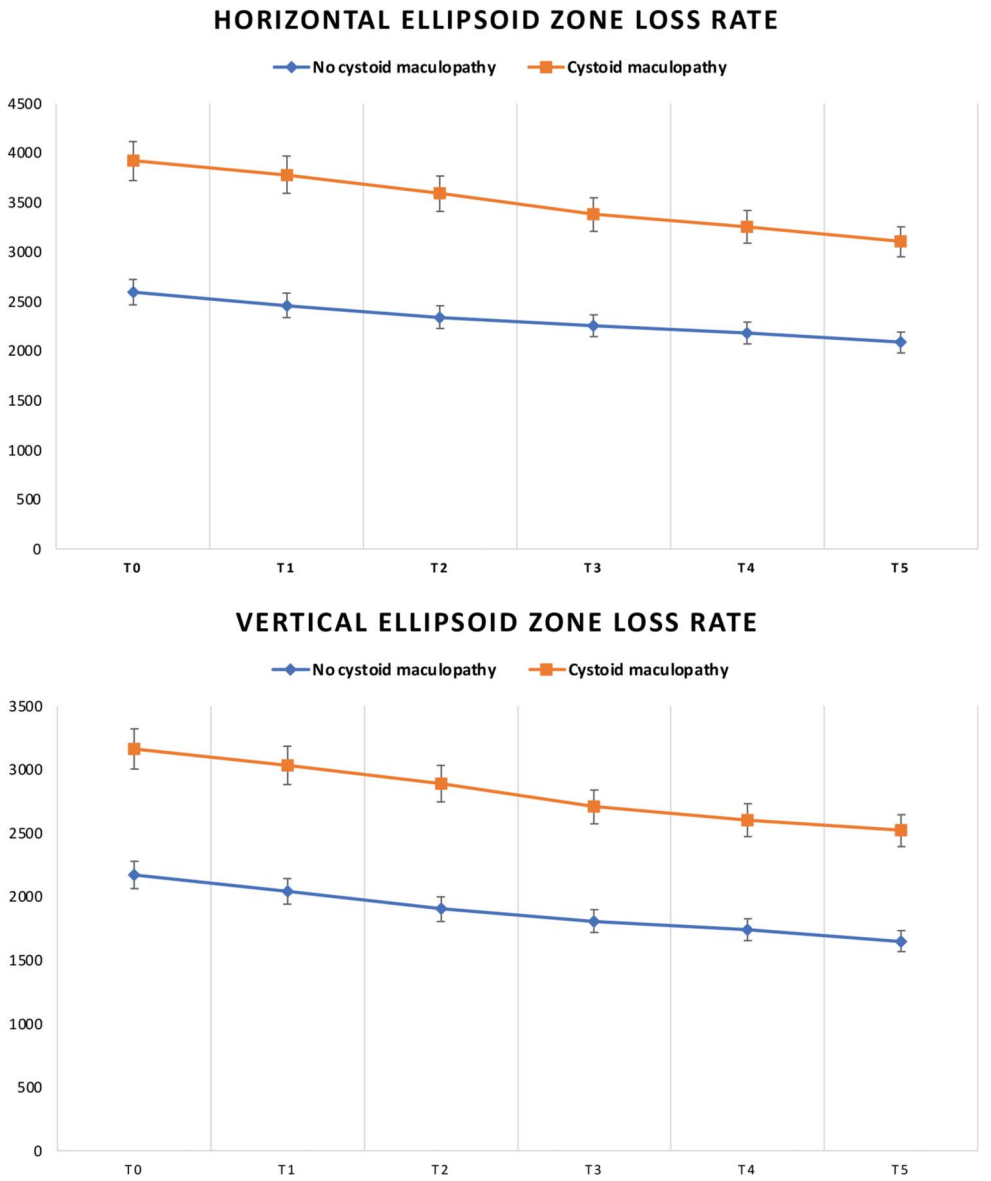

Fig. 1. EZ loss rate in RP with and without cystoid maculopathy.

regression started with the biggest EZ width, the EZ loss rate acceleration associated with CM regression caused the reaching of similar 5-year EZ width, about RP eyes with CM-P. Examples for different subgroups are reported in Figure 3.

Looking at RP eyes with CM-P, we registered a mean CM fluctuation of 21% (range 5%-34%) of total CMT value over the 5-year follow-up. We found no significant impact of CM fluctuation on the other considered metrics ($P > 0.05$).

The correlation analysis confirmed the association between CM and bigger EZ width (overall Tau–Kendall coeff. 0.522; $P < 0.001$). Moreover, we found a slight correlation between CMT and LogMAR BCVA (overall Tau–Kendall coeff. −0.312; $P = 0.02$).

Discussion

In this study, we evaluated the long-term impact of CM in eyes affected by RP related with *USH2A*. Our data confirmed the lack of a negative impact of CM both on visual acuity and EZ loss rate. However, if separately considering the behaviors of CM, we found that its complete regression was significantly associated with an acceleration of the EZ loss rate. Conversely, CM persistence and its fluctuations seem to have no relevant effect on the long-term outcome of RP eyes. The lack of a negative impact of CM on visual function has been already largely described,[3,7–11] associating the visual integrity with the status of the outer retinal structures more than

Table 2. Clinical and Imaging Metrics

Clinical and Imaging Metrics						
Parameter	No cystoid Maculopathy	Cystoid Maculopathy		*P*		
		CM-P	CM-R			
Group	1	2	3	1 versus 2	1 versus 3	2 versus 3
Number of eyes	25	10	15			
Age (years)	48 ± 14	51 ± 13	49 ± 8	>0.05	>0.05	>0.05
Gender (M/F)	12/13	5/5	7/8	>0.05	>0.05	>0.05
Spherical equivalent (diopters)	−3 ± 1	−3 ± 2	−3 ± 2	>0.05	>0.05	>0.05
LogMAR BCVA 0 (mean Snellen equivalent)	0.0 ± 0.1 (20/20)	0.2 ± 0.1 (20/32)	0.1 ± 0.1 (20/25)	>0.05	>0.05	>0.05
LogMAR BCVA 5 years (mean Snellen equivalent)	0.0 ± 0.2 (20/20)	0.2 ± 0.2 (20/32)	0.1 ± 0.2 (20/25)	>0.05	>0.05	>0.05
CMT 0 (μm)	285 ± 48	360 ± 41	405 ± 110	<0.001*	<0.001*	>0.05
CMT 5 years (μm)	258 ± 46	410 ± 42	418 ± 78	<0.001*	<0.001*	>0.05
CT 0 (μm)	278 ± 88	245 ± 52	275 ± 66	>0.05	>0.05	>0.05
CT 5 years (μm)	265 ± 89	223 ± 53	265 ± 56	>0.05	>0.05	>0.05
EZ horizontal 0 (μm)	2,568 ± 898	3,255 ± 712	4,220 ± 698	<0.001*	<0.001*	<0.001*
EZ horizontal 5 years (μm)	2057 ± 687	2,933 ± 335	3,142 ± 487	<0.001*	<0.001*	>0.05
EZ vertical 0 (μm)	2,168 ± 987	2,645 ± 695	3,399 ± 813	<0.001*	<0.001*	<0.001*
EZ vertical 5 years (μm)	1,645 ± 801	2,391 ± 387	2,537 ± 433	<0.001*	<0.001*	>0.05
TOT horizontal EZ loss (μm)	−508 ± 340	−322 ± 390	−1,048 ± 449	<0.001*	<0.001*	<0.001*
TOT vertical EZ loss (μm)	−523 ± 420	−253 ± 368	−825 ± 434	<0.001*	<0.001*	<0.001*
Mean horizontal EZ loss/year (μm/year)	−103 ± 68	−65 ± 78	−211 ± 88	<0.001*	<0.001*	<0.001*
Mean vertical EZ loss/year (μm/year)	−105 ± 82	−52 ± 72	−167 ± 87	<0.001*	<0.001*	<0.001*
ERM	8 (32%)	1 (10%)	6 (40%)	<0.001*	>0.05	<0.001*
HGB	5 (20%)	1 (10%)	5 (33%)	>0.05	>0.05	<0.001*

CMT, central macular thickness; ERM, epiretinal membrane.

the presence of CM. Interestingly, we found a minor CM impact on the visual function in the RP subgroup characterized by CM-P. In our previous study, we hypothesized the Müller cells dysfunction as the primary pathogenic source of CM in RP.[8] This may be explained considering that Müller cells govern the entire retinal fluid homeostasis.[12,13] However, foveal Müller cells are known to be characterized by real optical properties, favoring the concentration of the light stimuli on the photoreceptors.[12] On this basis, we might advance the hypothesis that visual acuity disturbances in RP with CM-P are more related with the partial loss of the optical and trophic properties of foveal Müller cells than a direct damage caused by the CM on macular photoreceptors.

Interestingly, we found a strong relationship between CM-R and EZ loss rate. In particular, the disappearance of CM has been associated with a faster reduction of the EZ band. It is known that the pathogenesis of CM is extremely complex, not being associated with the mere transudation of fluid in the extracellular space.[14] Although we have no molecular data to provide a definite interpretation of the described phenomenon, we may speculate about the possible transient loss of Müller cells–related trophic support on retinal cells occurring during CM regression, thus leading to the acceleration of EZ loss. Somehow, the homeostasis of the retina is restored after the CM disappearance, renormalizing the EZ loss rate.

Additional parameters, such as ERM and HGB, seem to have no significant impact on CM pathogenesis. On this basis, ERM might be interpreted as a reactive phenomenon occurring at the level of the vitreoretinal interface.[15] About HGB, the main pathogenic hypothesis regards the gliosis of the superficial capillary plexus.[9] The relatively higher prevalence of HGB in RP with CM-R might support the stressful nature of CM-R, leading to retinal cells damage. However, our data does not support a relevant role of HGB in the pathogenesis of CM.

We are aware that our study is potentially affected by several limitations, firstly related with the relatively low number of eyes. Moreover, a 5-year period may not be an adequate window of time to compare two groups with slow rate of disease. Longer follow-up periods could potentially yield different results. However, a strength point is that we performed this study

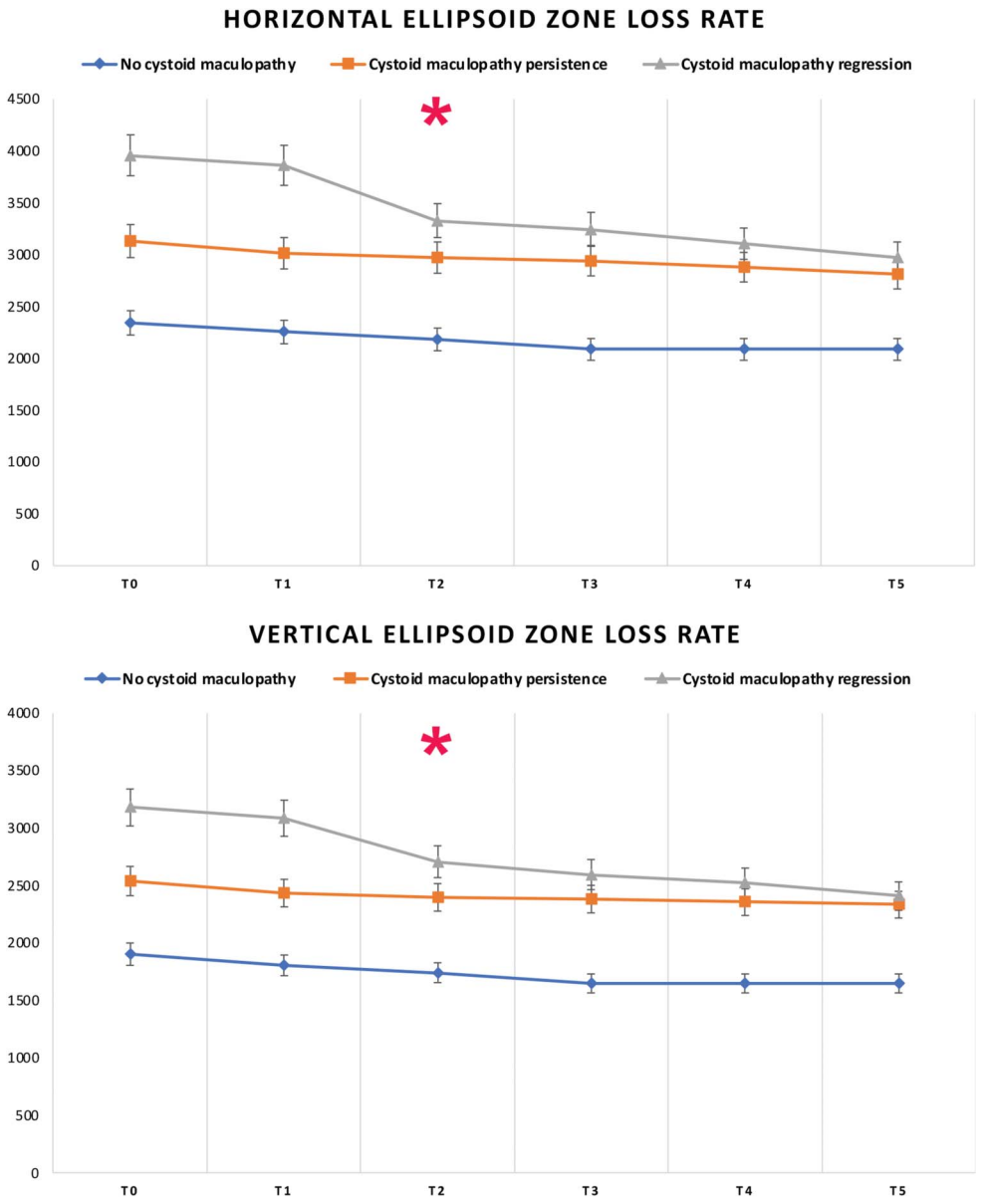

Fig. 2. EZ loss rate subgroups analysis. RP are divided in eyes without CM, eyes with CM persistence, and eyes with CM recurrence. The EZ loss rate acceleration secondary to CM regression is highlighted by red asterisks (*).

only considering *USH2A* RP to avoid possible effects caused by genotypes variability. Moreover, *USH2A* RP represents the most prevalent RP subtype in our country,[16–22] thus allowing to collect much more data. However, future studies should be focused on evaluating the impact of CM in RP caused by other pathogenic variants. Although we took several steps to guarantee high image quality, we are also mindful that all imaging techniques are prone to several artifacts.[23] Another limitation is related with the fact that we did not correct for axial length for EZ width measurements. However, the refractive range was similar among the three subgroups, thus reducing the possible influence of this limitation. In addition, provided several pathogenic speculations that will require molecular and histologic validations.

In conclusion, our study highlighted the long-term impact of CM in RP. Its regression is associated with the acceleration of EZ loss rate, whereas ME persistence and its fluctuations seem to have no relevant effect on the morphologic and functional outcome. Although our study cannot provide conclusive data, our findings support the hypothesis that CM pathogenesis is not characterized by active production of fluid but should be considered as the consequence of the loss of intraretinal fluid homeostasis. Therefore, we should refer to this as "cystoid maculopathy" instead of "cystoid macular edema." For this reason, although

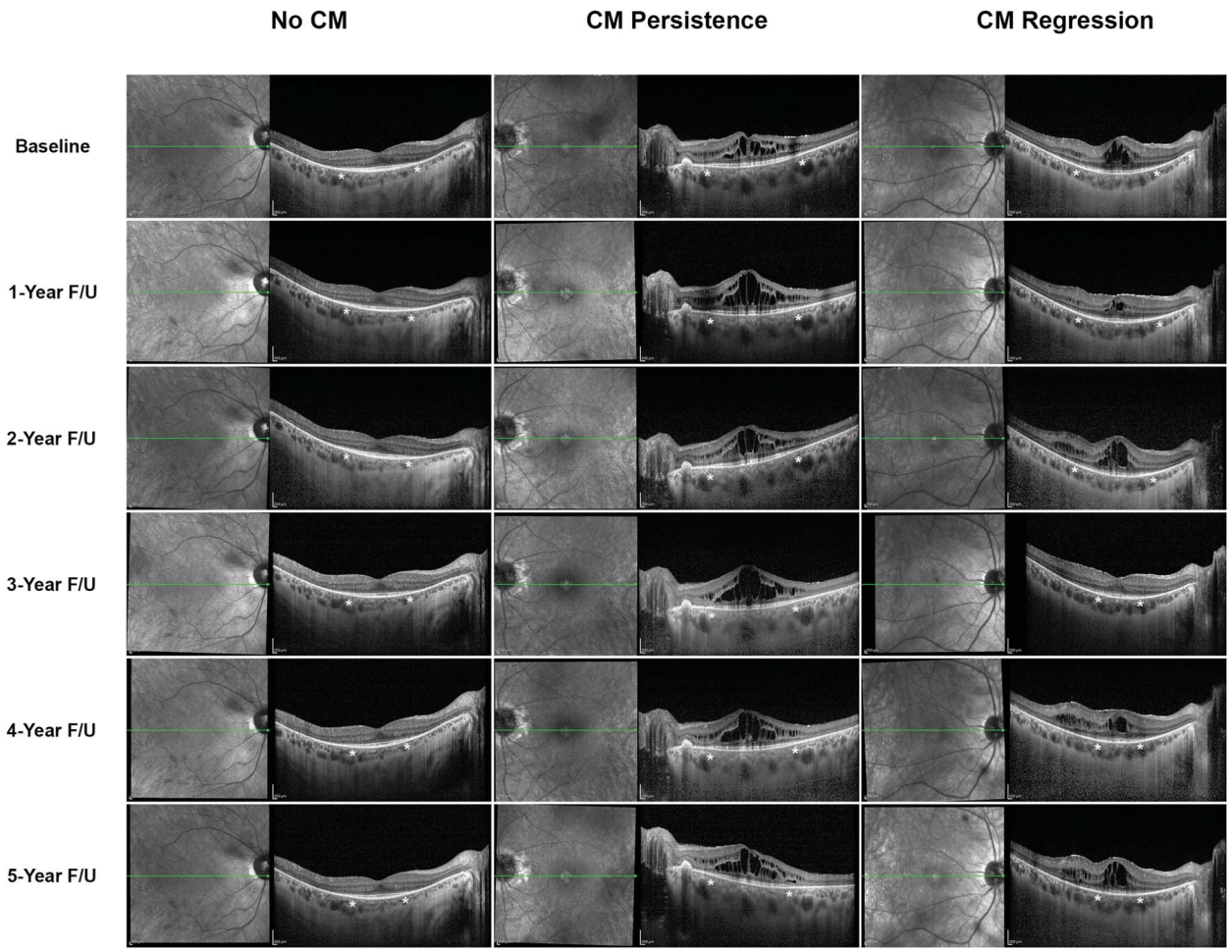

Fig. 3. The clinical impact of CM in RP. The first column shows a case of RP without CM. The EZ loss rate is slow and stable over the 5-year follow-up. The similar behavior is shown by the second column (RP with CM persistence). It is also possible to observe CM fluctuations not associated with EZ loss worsening. The last column shows a case of RP with CM recurrence. Also in this case, CM fluctuations have no remarkable effect on EZ loss rate. The CM regression (3-year follow-up) is associated with a remarkable worsening of EZ loss. After this time point, the EZ loss rate comes back to slow and stable progression.

conservative treatments such as topical or oral carbonic anhydrase inhibitors might have a role in the management of this condition, invasive and potentially dangerous approaches such as intravitreal injections or corticosteroids implants should be avoided. Further studies are needed to define CM clinical role in RP genotypes different from *USH2A* form.

Key words: retinitis pigmentosa, cystoid maculopathy, macular edema, EZ, BCVA.

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