Crumpled Impairment of Vision as a Sign of a Serious Eye Disease (and How to Manage It)[1]

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ABSTRACT

Introduction: Philosopher Thomas Reid, over two centuries ago, and later Richard Forster, described abnormal curvature of straight lines in vision, known as metamorphopsia, popularly said crumpled vision. Marc Amsler's grid, introduced almost a century afterward, remains a popular tool for evaluating this phenomenon. Advances in Optical Coherence Tomography (OCT) have allowed the study of retinal layers in vivo, shedding light on the morphological correlation of this symptom; Methods: This study utilizes fundus photography, OCT B-scans, and angiography to document macular findings, retinal layer morphology, and fluid dynamics. The fovea, a crucial region for detailed vision, is examined; Results: Crumpled vision, akin to distorted images in video-mapping, is linked to a convex deformation of the outer segments of photoreceptors due to subretinal fluid leakage, seen in macular detachment. Successful treatment at the initial stage is crucial, as long-term cases may paradoxically lose metamorphopsia but result in permanent central scotoma with profound visual loss; Case Studies: Four common diseases causing crumpled vision—Central serous chorioretinopathy, Exudative neovascular age-related macular degeneration, Diabetic maculopathy, and Vitreomacular traction—are presented. Successful treatments, including laser photocoagulation, anti-vascular endothelial growth factor injections, and pars plana vitrectomy with internal limiting membrane peeling, are detailed; Conclusion: Understanding the source of fluid leakage is crucial for tailored treatment. The study highlights successful interventions in treating crumpled vision, emphasizing the importance of early diagnosis and choice of right specific treatment method or their combinations. The "supermarket test" is proposed as a simple method to increase self-monitoring awareness among individuals with potential vision issues.

KEY WORDS

Crumpled Vision – Metamorphopsia – Amsler Grid – Optical Coherence Tomography – Retinal Layers – Macular Detachment – Outer Segments of Photoreceptors Layer – Central Serous Chorioretinopathy – Exudative Neovascular Age-Related Macular Degeneration – Diabetic Maculopathy – Vitreomacular Traction – Laser Photocoagulation – Anti-Vascular Endothelial Growth Factor Injection – Pars Plana Vitrectomy – Internal Limiting Membrane Peeling – Supermarket Test

Philosopher Thomas Reid was the first one who described two hundred and sixty years ago an abnormal curvature of straight music lines when he looked with the right eye only.[2] One hundred years later Richard Forster published several case reports about crumpled vision (which is the meaning of a Greek word „metamorphopsia").[3] Marc Amsler's grid for the evaluation of this problem, which appeared in the literature after another almost one hundred years, is very popular till today, fig. 1.[4] But the morphological correlation of this symptom remained unclear until the era of Optical coherence tomography (OCT), which enabled the study of retinal layers in vivo, just 30 years ago.[5]

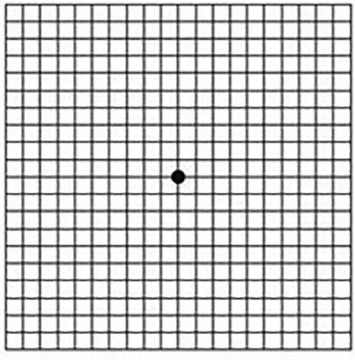

Fig. 1. Amsler grid.

An example of how to simulate a patients' problem is video-mapping performed on an irregular building wall, fig. 2.

Fig. 2. Projection of a hockey goal gate with a puck to the front wall with columns of the National Museum building in Prague.[6]

Distortion of the image makes it difficult to see details. It may be noticed how the inner yellow ring of the puck is discontinued.

The "macula" in the eye is the most efficient retinal part regarding image sharpness and resolution. In this article, macular findings are documented by fundus photography (color and red free (RF) type), retinal layers morphology by

OCT B-scans, and fluid dynamics by angiography, fig. 3. OCT B-scans are exported by X:Y axis ratio 1:2 or 1:3 depending on the purpose of evaluation.

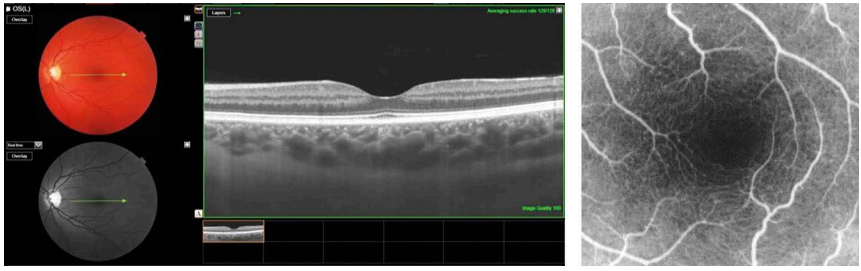

Fig. 3. Left eye with normal findings: images of the posterior pole of the back of the eye (color and RF) – left, Retinal OCT B-scan (the plane of cross-section is situated within the green arrow on the fundus picture) – middle, angiography showing normal perfusion of retinal vessels and capillary free zone in the center of the macula (dark area) – right.

The fovea is the 1,5 mm wide circular depression in the macular center, the site of the most detailed vision, fig. 4. On its floor – foveola – we find only single neuronal cell layer of photoreceptors. And because it is a part of the central nervous system, in case of damage, it can't regenerate. On the OCT B-scan we can recognize all layers described histologically. In this article, however, we focus on the subcellular one: the outer segments of photoreceptors, which is the site of image projection and its perception. Normally, it is straight and parallel with retinal pigment epithelium (RPE).[7]

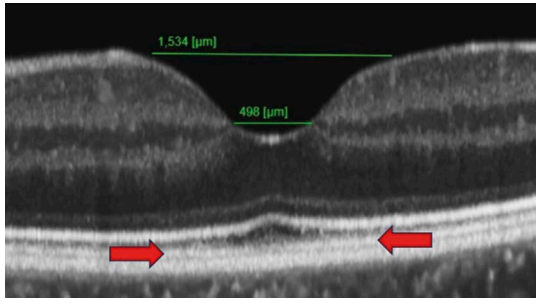

Fig. 4. Normal OCT B-scan: Extent of the fovea (longer green line) and capillary free zone (shorter green line, compare with angiography image on the fig. 3), red arrows at the level of second hyperreflective band from the choroid (in the bottom) = outer segments of photoreceptors layer.

If we simplify the crumpled vision pathophysiology, the previously mentioned phenomenon of distorted image by columns during video-mapping has its similarity on the pathologically convex shape of the outer segments of the photoreceptors layer,

fig. 5. It develops due to subretinal fluid leakage (so-called macular detachment).[8] Although RPE remains flat, outer segments of the photoreceptors layer are elevated.

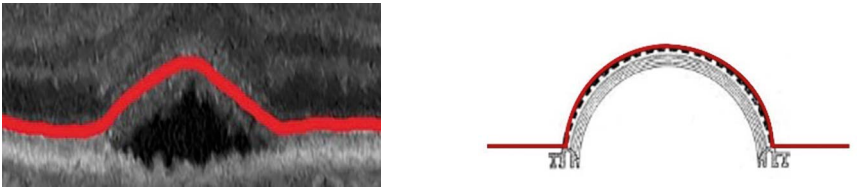

Fig. 5. Pathologic OCT B-scan in detail: red line = convex deformation of outer segments of the photoreceptors layer due to its detachment from RPE in the fovea (left). Animation of a cross-section of the building wall with the semi-column: red line = its surface with convex deformation in the center (right).

Metamorphopsia comes together with slightly decreased vision as the metabolism of photoreceptors is broken due to lost contact with RPE. When the disease lasts a long time, metamorphopsia paradoxically disappears – it is when a patient has deep visual loss with a permanent absolute central scotoma. It means that this sight-threatening condition is treatable only at an initial stage.

The four most frequent diseases leading to crumpled vision are presented through case reports of our patients as examples. They are: Central serous chorioretinopathy (CSCR), Exudative neovascular age-related macular degeneration (ENAMD), Diabetic maculopathy (DM) and Vitreomacular traction (VMT). All of them had the same finding of macular detachment, but the difference between them is in the source of fluid leakage. Our treatment (Tx) in all cases was successful, leakage stopped, and residual fluid resorbed – macula dried. But the recurrence of macular detachment is likely to develop if the state-of-art Tx method is not causal.

The source of fluid in CSCR is in the lowest level (thickened choroid), fig. 6. Retinal detachment is due to imbalance of the outer blood-retinal barrier. It is found mostly in men from 20 to 50 years of age. Tx, which is symptomatic, consists of laser photocoagulation, which blocks up the RPE defect. An angiography is needed to exactly localize it. An angiography is performed using a fluorescein dye injected intravenously and the macula is captured with special filters of retinal camera (so called Fluorescein angiography, FA).[7]

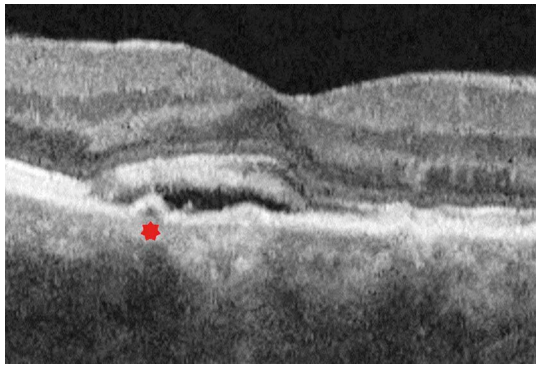

Fig. 6. OCT B-scan in CSCR: Thickened choroid is the source of leakage (see the red asterix under the RPE).

Patient 1 was a 40-year-old man with CSCR. If we look to his right eye FA image at presentation, there is a hot spot at 11 o'clock position regarding to the foveal center. He had metamorphopsia due to macular detachment, which is visible on the OCT B-scan, fig. 7. One month after performing laser at the site of the hot spot metamorphopsia disappeared and the macula was dry.

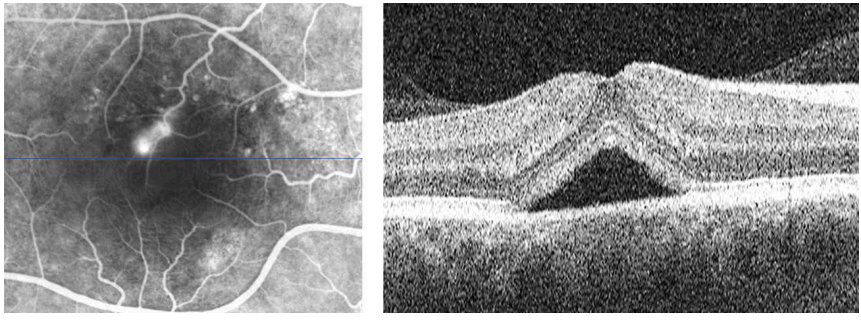

Fig. 7. Patient 1 at presentation: FA image (left), OCT B-scan – the plane of cross-section is situated within blue line on FA image (right).

We successfully treated two recurrences, 4 and 8 years thereafter. 3rd recurrence developed after another 18 months, fig. 8. There were three scars at 11, 1 and 2-3 o'clock on FA. Hot spot at 10 o'clock position was managed by the laser again. Metamorphopsia disappeared together with a shallow macular detachment within a month and the patient had no problem.

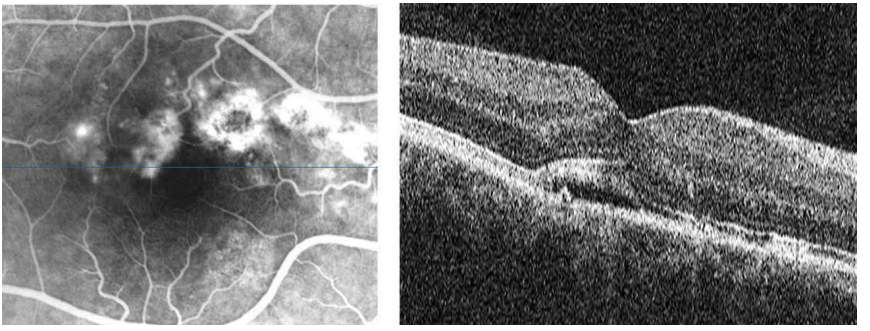

Fig. 8. Patient 1 after 8 years of the follow-up: FA image (left), OCT B-scan – the plane of cross-section is situated within blue line on FA image (right).

At the last visit, 6 months after the last recurrence, we see quiet scars on RF image and normal macular configuration on OCT B-scan, fig. 9.

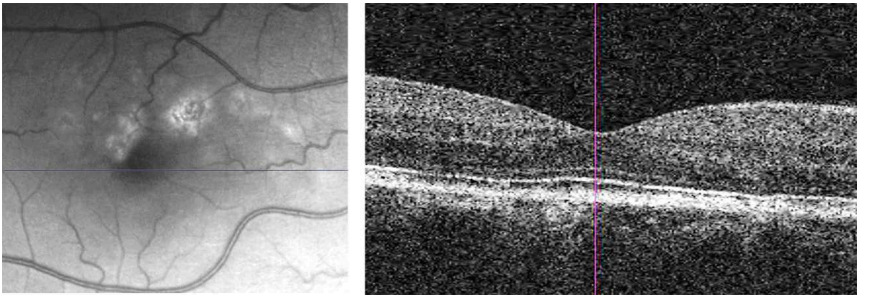

Fig. 9. Patient 1 at the last visit: RF image (left), OCT B-scan – the plane of cross-section is situated within blue line on RF image (right), violet vertical line = center of the foveola.

The source of fluid in ENAMD is so-called macular neovascularization (MNV), fig. 10. The retinal detachment is also due to an imbalance of the outer blood-retinal barrier. ENAMD is found after 50 years of age.[7] Tx, which is symptomatic, consists of injections of anti-vascular endothelial growth factor (anti-VEGF) intravitreally. The patient is seen each month and repeated injection administration is performed, when needed.[9] Each injection has possible side effects, mainly intraocular infection. If angiography excludes subfoveal MNV extent, it can be burned by a laser.[10] It is the only available possibility to decrease the number of needed injections.[11]

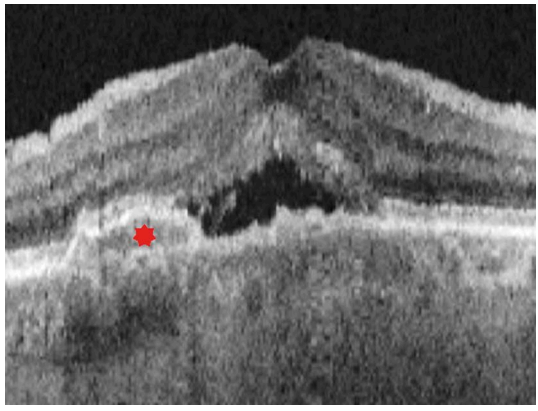

Fig. 10. OCT B-scan in ENAMD: MNV is the source of leakage (see the asterix above the choroid).

Patient 2 was a 75-year-old woman with ENAMD. If we look to her right eye OCT-Angiography image (OCTA, en face technique of high-resolution OCT) there is extrafoveal MNV. Because it is not visible during a laser, it was needed to prepare a Tx plan with an overlay technique to RF image. Patient is instructed not to looking for a laser beam, because the shot to the center would lead to immediate and permanent visual loss. On the second RF image we see posttreatment appearance with confluent light laser spots. Their extent from the fovea is 0,5 mm, fig. 11.

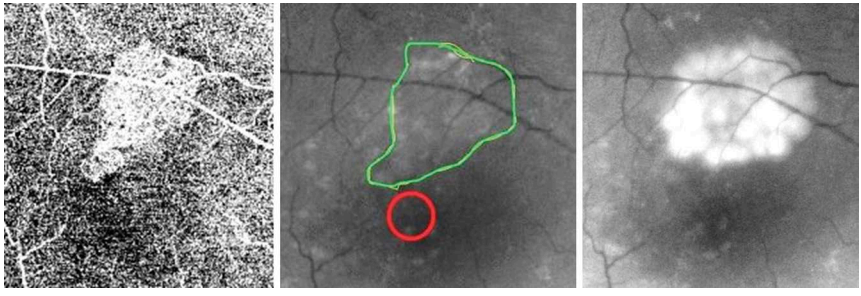

Fig. 11. Patient 2: OCTA at presentation (left), Tx plan (red circle is the fixation area which must not be coagulated, area depicted in green is an overlayed extent of the MNV) in RF image (middle), posttreatment RF image (right).

After the laser, an anti-VEGF injection was administered to prevent the recurrence. Metamorphopsia disappeared within a month. The macula dried as it is seen from comparison of OCT B-scans. MNV is no longer detectable on OCTA image compared to the pre-treatment one 7 months after the initial Tx, fig. 12.

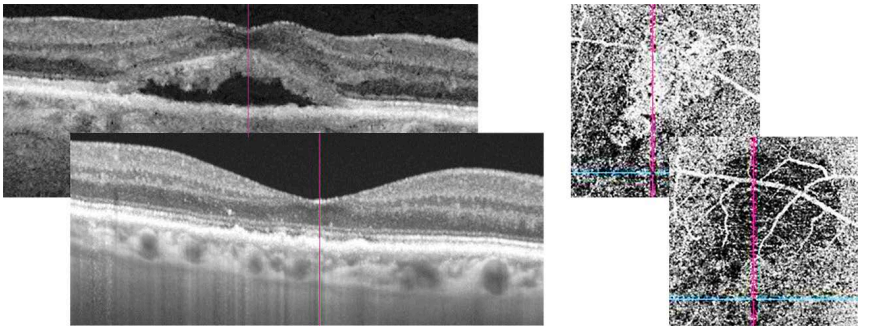

Fig. 12. Patient 2: OCT B-Scan at presentation (left above) and at the last visit (left below) – the planes of cross-section are situated within blue horizontal line on OCTA images, violet vertical line = center of the foveola, OCTA images at presentation (right above) and at the last visit (right below), crossing of the lines = fixation point.

In DM, the fluid leaks from the intraretinal microaneurysms (MAs), which is an eye manifestation of systemic diabetic microangiopathy, fig. 13. The retinal detachment is due to an imbalance of the inner blood-retinal barrier. It is found in patients after at least of 10 years of duration of diabetes mellitus. Tx, which is symptomatic, consists of injections of anti-VEGF intravitreally combined with direct laser of MAs. FA is needed to localize precisely the leaking MAs and differentiate them from inactive ones and from hemorrhages.[7]

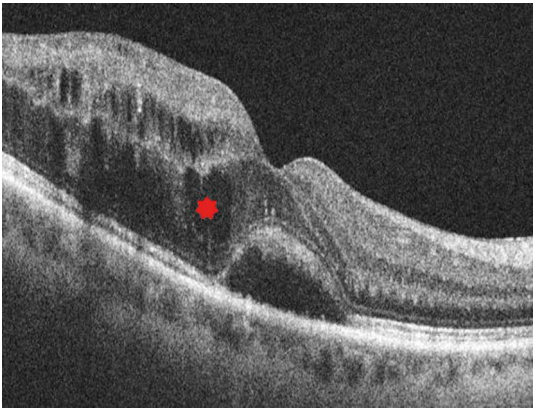

Fig. 13. OCT B-scan in DM: MAs are the source of the leakage (see the asterix above the outer segments of photoreceptors layer).

Patient 3 was a 69-year-old man with diabetic maculopathy and diabetes lasting for 17 years. If we look to his right eye RF image, there are a lot of dark spots. It would not be possible to decide which one should be treated by laser without FA, on which only MAs are hyperfluorescent (light hot spots with a maximal size of 0,1 mm), fig. 14.

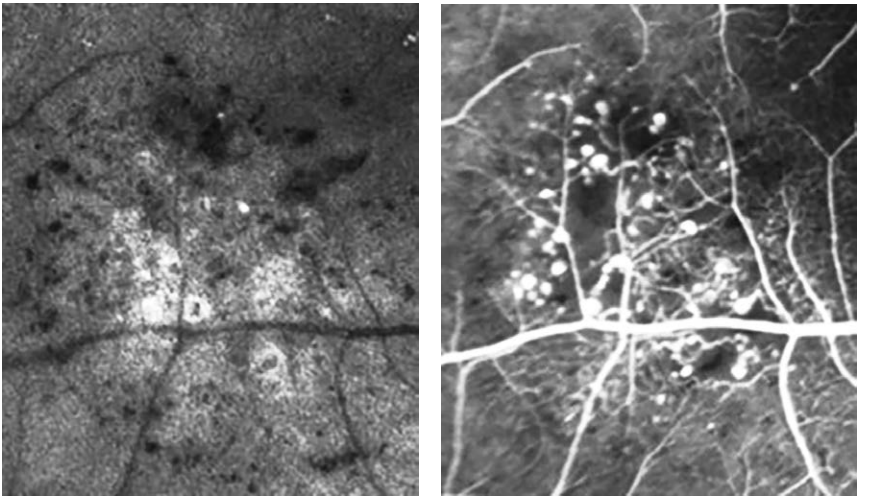

Fig. 14. Patient 3 at presentation: RF image (left), FA image (right).

2 months after anti-VEGF injection intravitreally and laser, the patient had no metamorphopsia and the macula became dry for the next 4 years, as can be seen on the last OCT B-scan compared to the pre-treatment one, fig. 15.

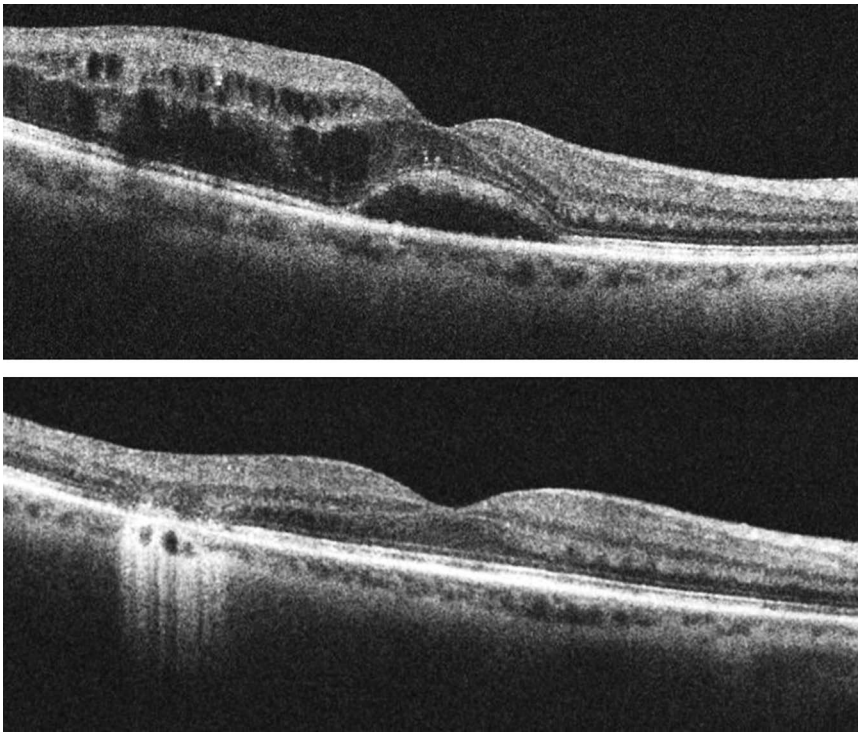

Fig. 15. Patient 3: OCT B-scans before Tx (above), at the last visit (below).

In vitreomacular traction (VMT), fluid leakage develops due to excessive tractional force at the level of the vitreoretinal interface. The normal process of posterior vitreous cortex (VC) detachment may become anomalous if VC adhesion to the macula is too firm, fig. 16. The disease is found between 60 and 80 years of age. Tx is causal and consists of the pars plana vitrectomy (PPV). During the operation, the traction is resolved by surgical posterior vitreous detachment. Internal limiting membrane (ILM) peeling is also performed during this operation in cases when an epiretinal membrane is present.[12]

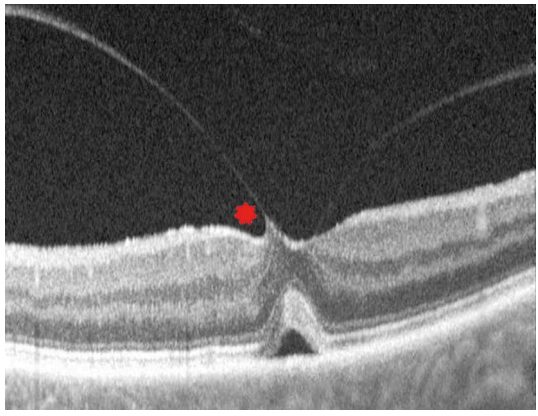

Fig. 16. OCT B-scan in VMT: Traction of the retina by anomalous firm adhesion of the VC (see the asterix above the retina).

Patient 4 was a 75-year-old man with VMT. One month after PPV the macula was dry, and the patient no longer has metamorphopsia, fig. 17.

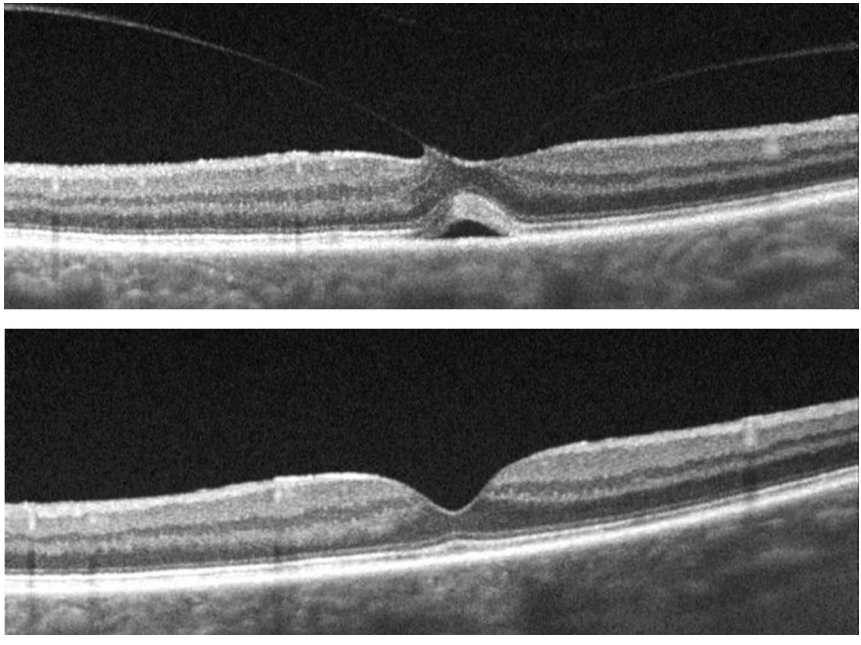

Fig. 17. Patient 4: OCT B-scan before (above) and after PPV (below).

Because the posterior vitreous cortex is almost invisible, it is demarcated by a small amount of suspension of triamcinolone crystals during PPV. Then a vitrectomy probe is used to cut and suck out material from the eye including VC. ILM is also invisible, so we color it with a brilliant blue dye. It is peeled off by a tiny forceps atraumatically, fig. 18. All instruments are thinner than 1 mm, and they are exchanged through the implanted ports. Such an operation enables us to perform a sutureless operation without general anesthesia as an outpatient procedure.[12]

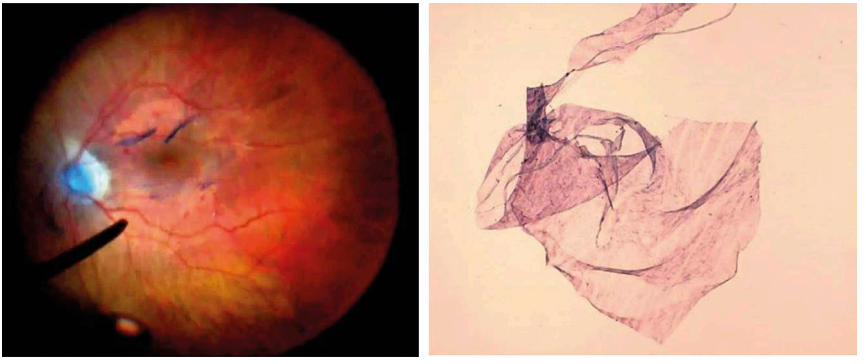

Fig. 18. ILM: Fundus image captured during PPV (blue-colored ILM is best visible on its rolled margins after beginning of peeling. The probe in the surgeon's left hand works as the endo-illumination), left. Histology image of the extracted piece of ILM, enlargement ×50. Reprinted from CIHELKOVÁ, I. and SOUČEK, P. Atlas of Macular Diseases[7], right.

There are different tools in daily life, how to perform Amsler grid: lines on the street, windows of the buildings, tiles in the bathroom, people's faces, letters when reading. But if looking with both eyes open, the brain suppresses the eye with a worse image. This is why people often overlook that the macular detachment on their first diseased eye has already developed. Then in a real world a patient comes to the medical office only after the disease is bilateral with untreatable condition on the worse eye. Method, how to increase poor rate of self-monitoring among those „healthy" people is simple: closing the fellow eye.

So, we propose the following "supermarket test". If one would look through an American "OK gesture", he or she probably close the fellow eye automatically. In addition, circle made by fingers focuses image to the fovea, fig. 19.

Fig 19. Supermarket test: normal image (left), crumpled vision with "damaged shopping cart, cut bananas and tomatoes or twisted baguettes" (right). Images courtesy: Centrum klinické oftalmologie s.r.o.

Four hundred years ago Comenius published "The Labyrinth of the World and the Paradise of the Heart".[13] According to the content of this article we would like to paraphrase an observation in this masterpiece: „The difference is in vision. It depends on the inner disposition … how one perceives an image, whether regular or crumpled".

NOTES

1 Part of this article was presented at the Comenius Academic Club Conference in Naarden, which took place in June 2023. Authors do not have a financial interest/arrangement or affiliation with one or more organizations which could be perceived as a real or apparent conflict of interest in the context of the subject of this article. Acknowledgements: staff members of our private eye clinic and referring physicians.

2 Thomas REID, An Inquiry into the Human Mind on the Principles of Common Sense, Edinburgh-London 1810 (6th ed.) The first edition was published in 1764.

3 Richard FÖRSTER, Ophthalmologische Beiträge, Berlin 1862.

4 Marc AMSLER, Earliest Symptoms of Diseases of the Macula, in British Journal of Ophthalmology, vol. 37 1953, pp. 521–537.

5 Eric A. SWANSON, Joseph A. IZATT, Michael R. HEE, et al. In Vivo Retinal Imaging by Optical Coherence Tomography, in Optics Letters, vol. 18 1993, pp. 1864–1866.

6 https://www.tyden.cz/rubriky/relax/ostatni/video-videomapping-rozzaril-budovu-narodniho-muzea_501118.html, accessed on-line on 1/APR/2023.

7 Ilona CIHELKOVÁ and Petr SOUČEK, Atlas makulárních chorob, Atlas of Macular Diseases, Prague 2005. (1st edition)

8 J. Donald M. GASS, Stereoscopic Atlas of Macular Diseases, St. Louis 1997. (4th ed.)

9 THE CATT RESEARCH GROUP, Ranibizumab and Bevacizumab for Neovascular Age-Related Macular Degeneration, in The New England Journal of Medicine, vol. 364 2011, pp. 1897–1908.

10 Petr SOUČEK, Jarmila BOGUSZAKOVÁ, Zdeňka GAJDOŠÍKOVÁ, et al. Diagnostika, sledování a laserová léčba klasické formy chorioideální neovaskularizace u pacientů s věkem podmíněnou makulární degenerací. [Diagnosis, Monitoring, and Laser Therapy

of the Classic Form of Choroidal neovascularization in patients with age-related macular degeneration], in Československá oftalmologie, vol. 53 1997, pp. 94–100.
11 Daniel WILL, Alexandra WILL, Combined Anti-VEGF Injections and Thermal Laser Photocoagulation for Peripapillary and Extrafoveal Choroidal Neovascular Membranes in Age-Related Macular Degeneration, in Investigative Ophthalmology & Visual Science, vol. 63 2022, p. 3804 – F0225.
12 Petr SOUČEK, Ilona CIHELKOVÁ, Josef ŠACH, Choroby vitreoretinálního rozhraní, II. část. [Diseases of the Vitreoretinal Region, Part II], in Česká a slovenská oftalmologie, vol. 60 2004, pp. 373–377.
13 http://www.labyrint.cz/en/chapter-22, accessed on-line on 5/APR/2023.