Université Paris Cité

Hôpital Lariboisière
Hôpital Saint Louis
AP-HP.Nord

HÔPITAL FONDATION
Adolphe de ROTHSCHILD
LA RÉFÉRENCE TÊTE ET COU

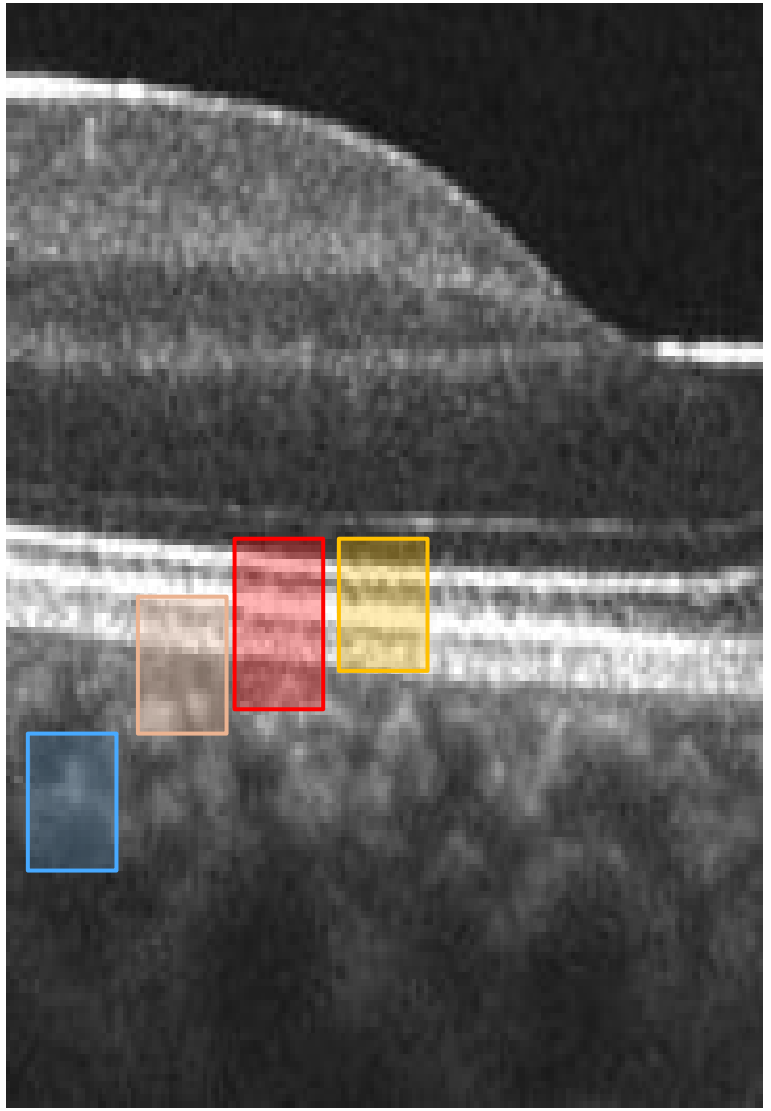
Multiple Evanescent White Dot Syndrome
MEWDS

Alain Gaudric

Service d'Ophtalmologie
Hôpital Lariboisière

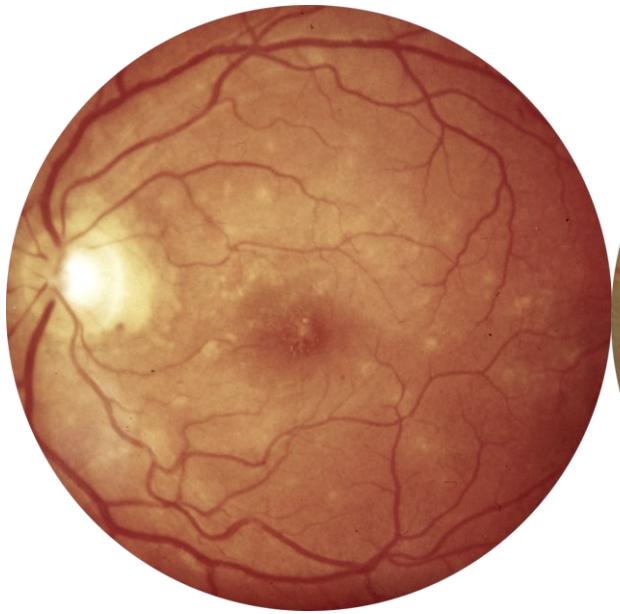
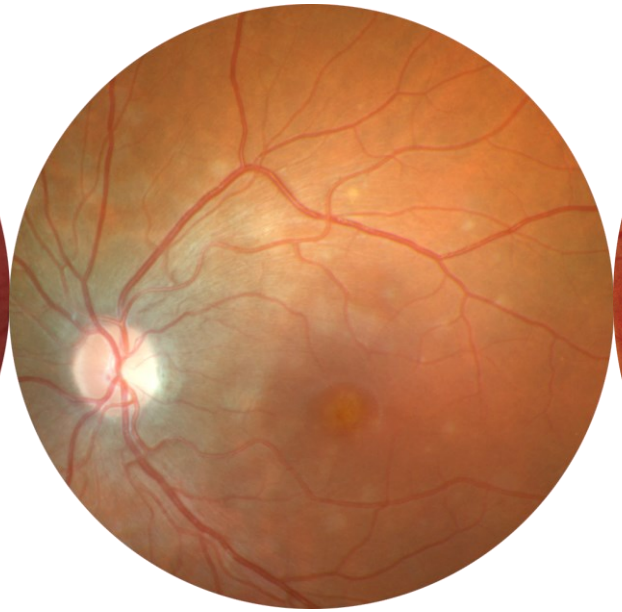
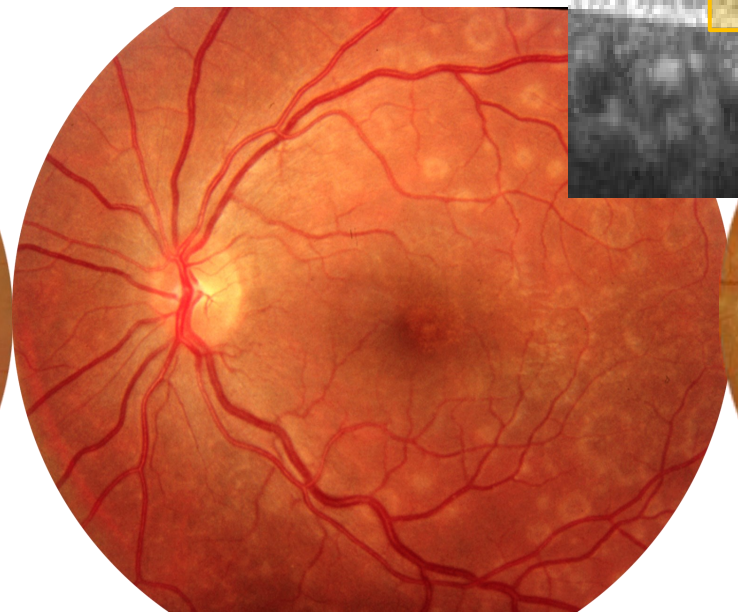
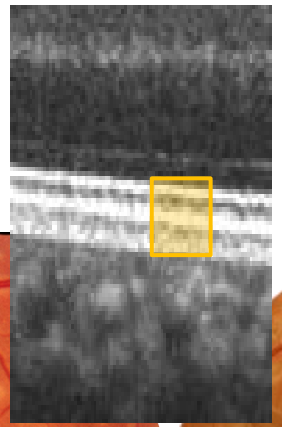
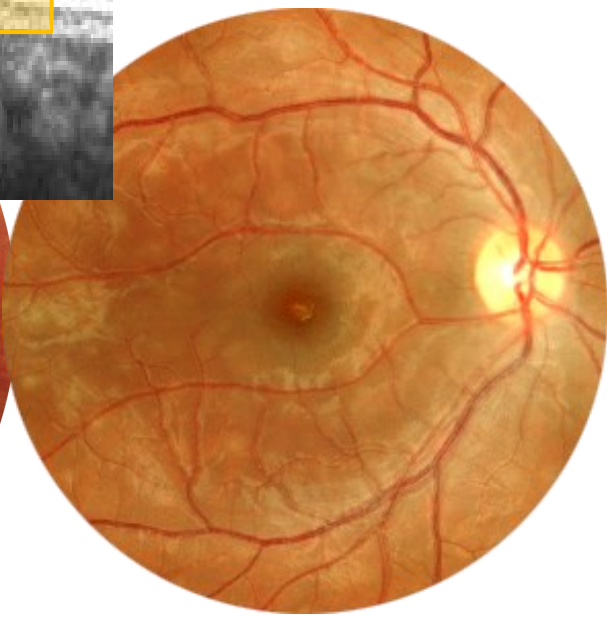
Essai de classification des taches blanches inflammatoires selon leur localisation

- Choroïde moyenne
  - Birdshot chorioretinopathy
    - mais aussi atteinte secondaire possible de l'épithélium pigmentaire et de la rétine
  - Sarcoïdose
    - mais aussi atteinte possible de la rétine, du NO , du SA
- Choroïde interne
  - Choroïdite ponctuée interne et choroïdite multifocale
- Choriocapillaire
  - Epithéliopathie en plaques et choroïdite serpigineuse
- Epithélium pigmentaire
  - Multiple Evanescent White Dot Syndrome



Multiple Evanescent White Dot Syndrome

- Le MEWDS atteint le complexe EP/PR sans atteinte de la choroïde.
- Les taches jaunes sont relativement discrètes et disparaissent en une à 2 semaines
- L'ICG est la clé du diagnostic

Multiple Evanescent White Dot Syndrome

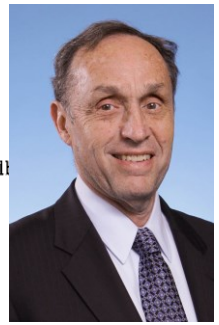
- Description par Lee Jampol, 1984
- femmes , 75 %%
  - jeunes , 25-30 ans
  - sans antécédents
- symptomes
  - baisse d ’AV
    - revenant à la normale en 6 semaines
  - photopsies
  - scotome paracentral
- champ visuel
  - élargissement de la tache aveugle
- ERG :
  - reduction de l ’onde a de l ’ERG

Multiple Evanescent White Dot Syndrome

I. Clinical Findings

Lee M. Jampol, MD; Paul A. Sieving, MD, PhD; David Pugh, MD; Gerald A. Fishman, MD; Howard Gil

Jampol LM, et al. Multiple Evanescent White Dot Syndrome: I. Clinical Findings. Arch Ophthalmol. 1984;102(5):671–674



Lee Jampol

INDOCYANINE GREEN ANGIOGRAPHIC ASPECTS OF MULTIPLE EVANESCENT WHITE DOT SYNDROME

AKIRA OBANA, MD, MASAYO KUSUMI, MB, TOKUHIKO MIKI, MD

Retina. 1996;16(2):97-104.

EXPANDED CLINICAL SPECTRUM OF MULTIPLE EVANESCENT WHITE DOT SYNDROME WITH MULTIMODAL IMAGING

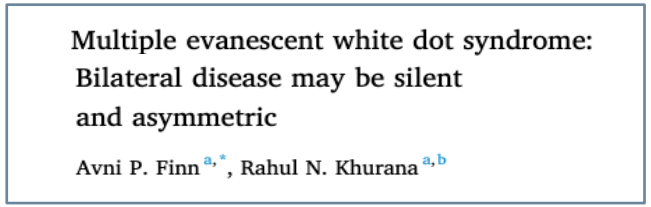
MARCELA MARSIGLIA, MD, PHD,*† ROBERTO GALLEGO-PINAZO, MD, PHD,‡ EDUARDO CUNHA DE SOUZA, MD,§ MARION R. MUNK, MD,¶** SUQUIN YU, MD,†† SARAH MREJEN, MD,‡‡ EMMETT T. CUNNINGHAM, JR., MD, PHD,§§ BRANDON J. LUJAN, MD,¶¶ NAOMI R. GOLDBERG, MD, PHD,*** THOMAS A. ALBINI, MD,††† ALAIN GAUDRIC, MD,‡‡‡ CATHERINE FRANCAIS, MD,§§§ RICHARD B. ROSEN, MD,¶¶¶ K. BAILEY FREUND, MD,*†**** LEE M. JAMPOL, MD,¶ LAWRENCE A. YANNUZZI, MD*†****

RETINA. 2016;36(1):64-74..

MEWDS

- Lésions récentes :
  - multiples taches , jaunes , au niveau de l ’EP ou de photorécepteurs,
  - moyennement opaques,
  - au pole pôstérieur et moyenne périphérie
  - flou papillaire
  - aspect granité, rouge-orange de la macula
  - quelques cellules dans le vitré
- Angiographie
  - fluo
    - iso ou hyperfluorescence discrète précoce et tardive.
    - hyperfluo papillaire
  - ICG
    - nombreuses taches hypofluo tardives

- Le MEWDS est habituellement unilatéral


Multiple evanescent white dot syndrome: Bilateral disease may be silent and asymmetric

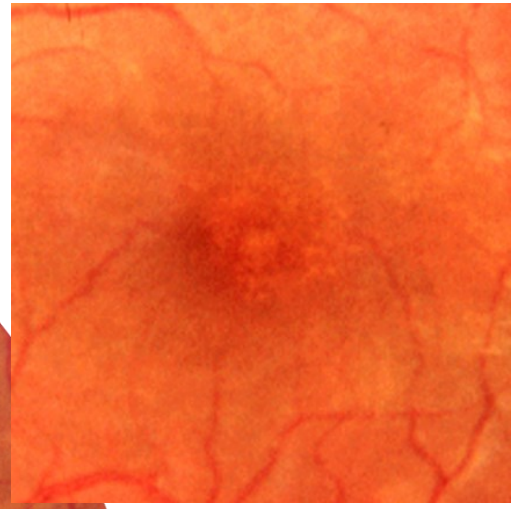
Avni P. Finn [a,*], Rahul N. Khurana [a,b]

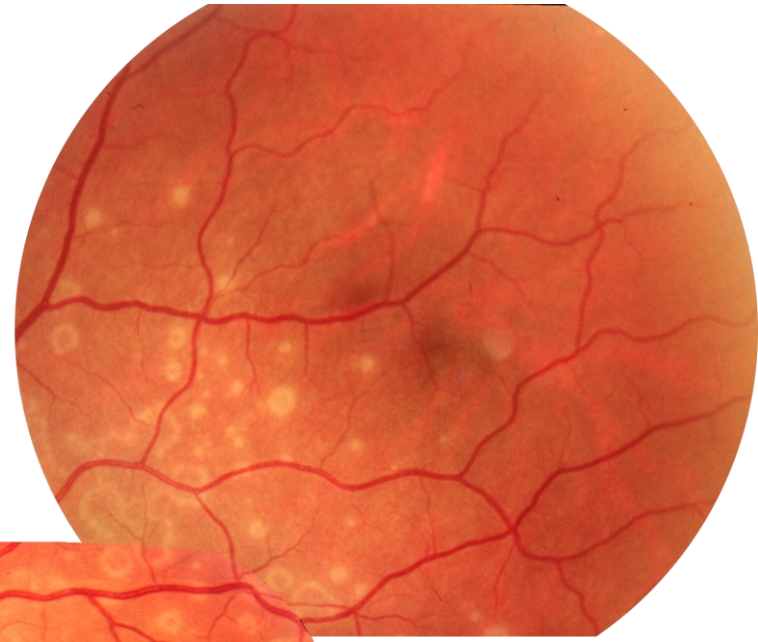
Am J Ophthalmol Case Reports. 2020;21:101004.

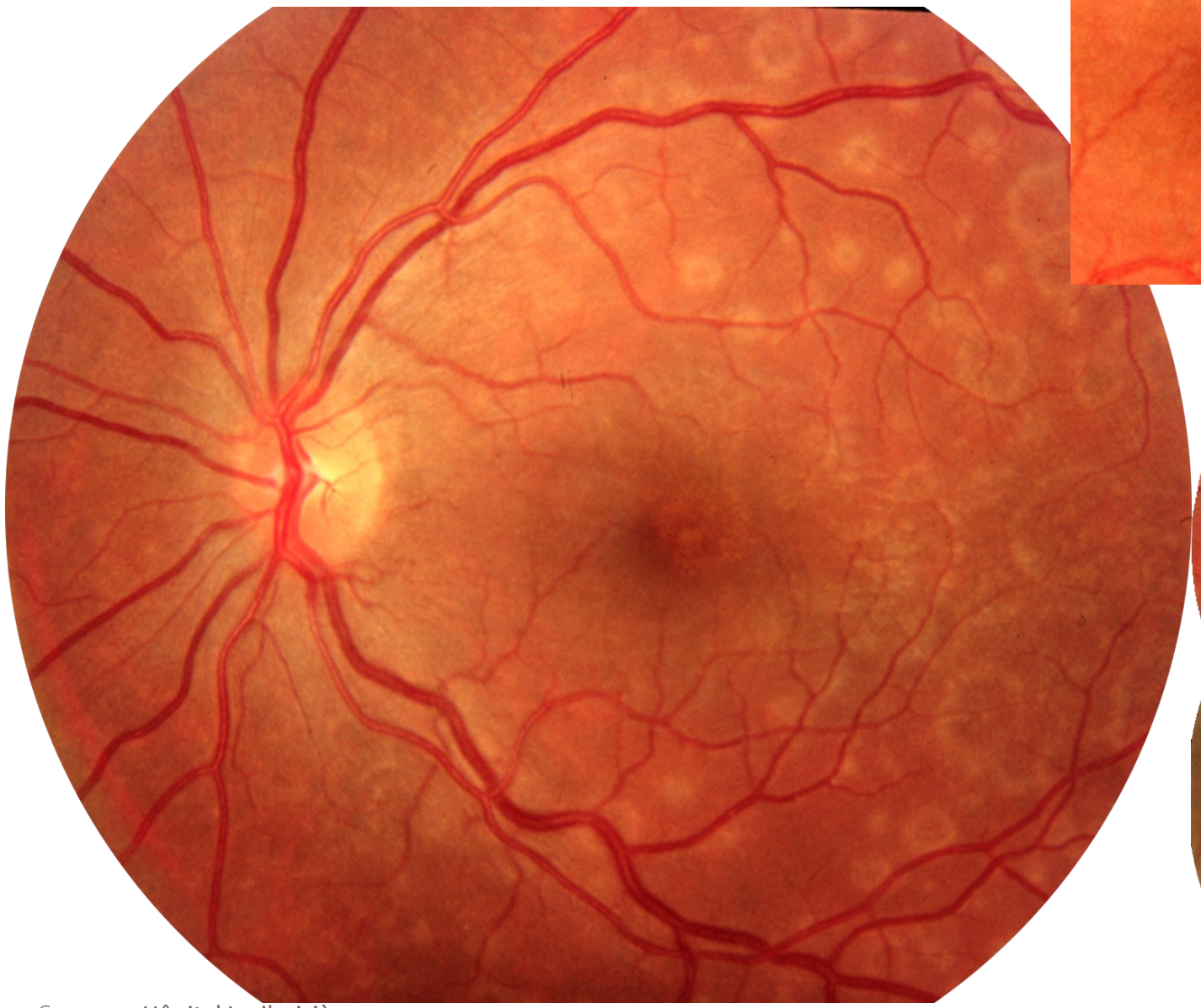
  - Mais des cas bilatéraux ont été décrits
  - parfois seulement révélés par les clichés en AF

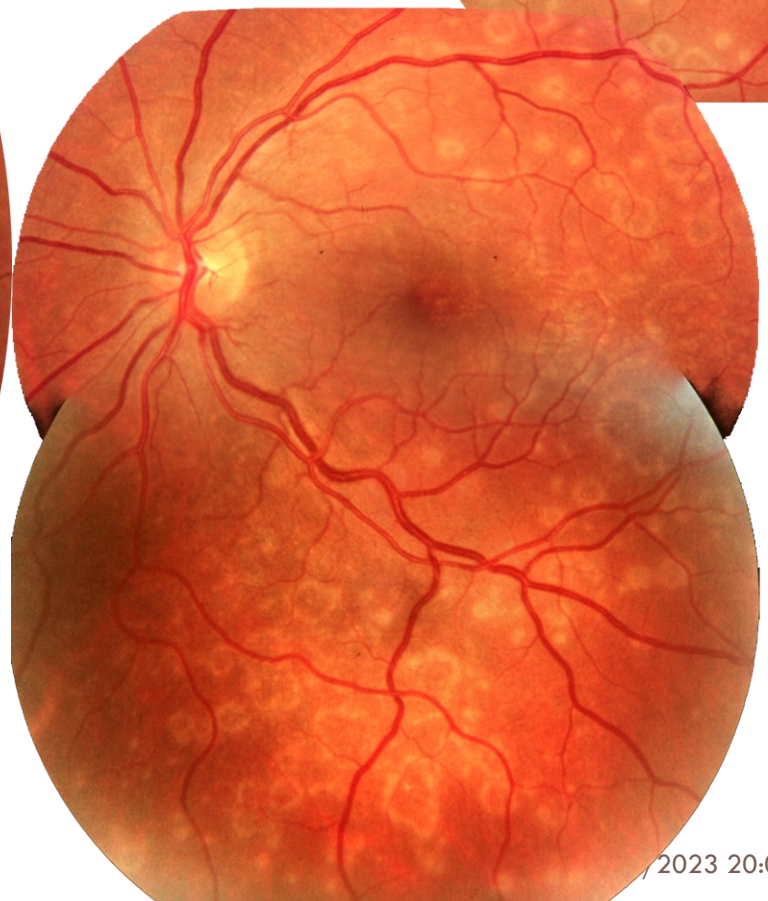
MEWDS

Granité maculaire





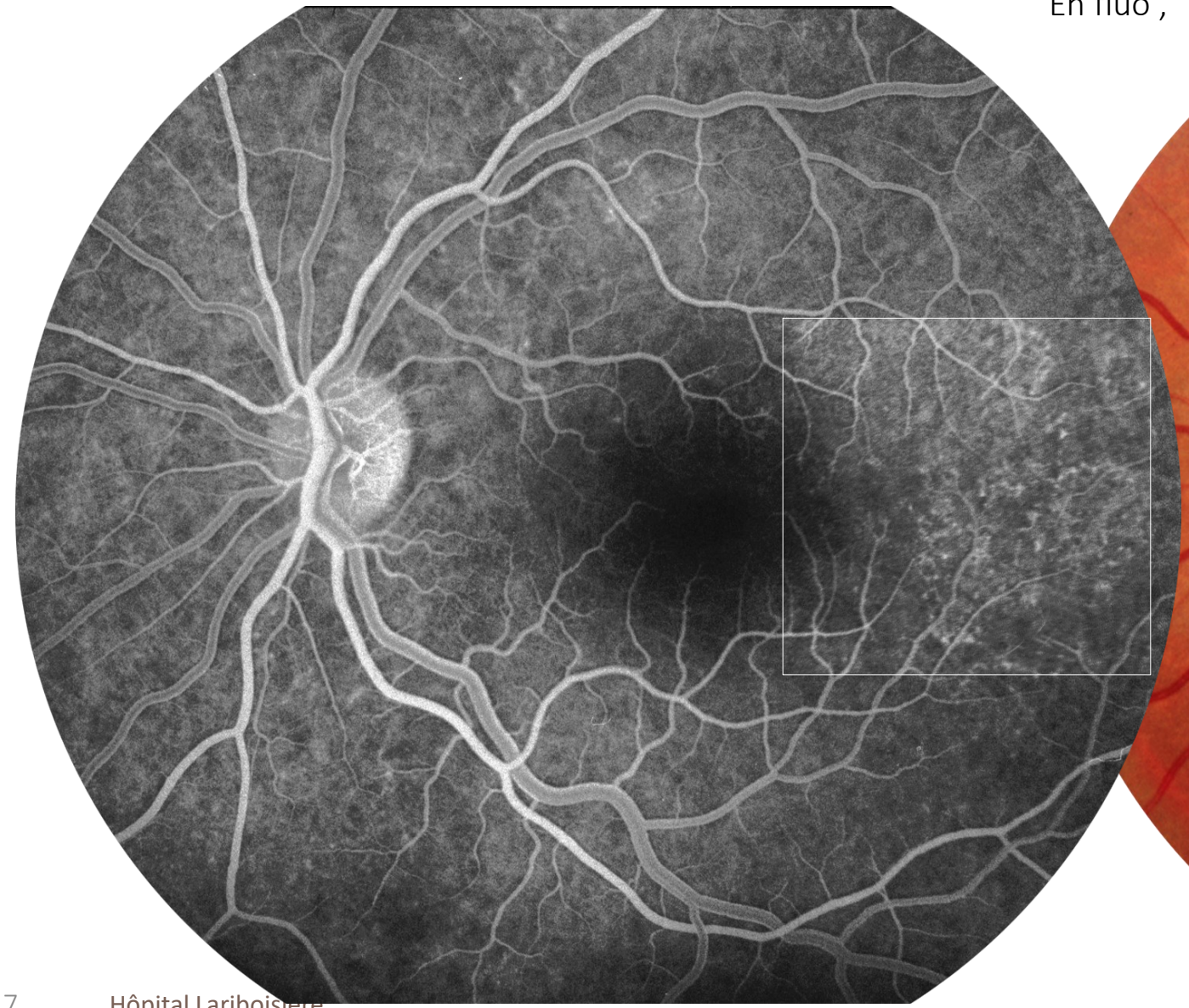


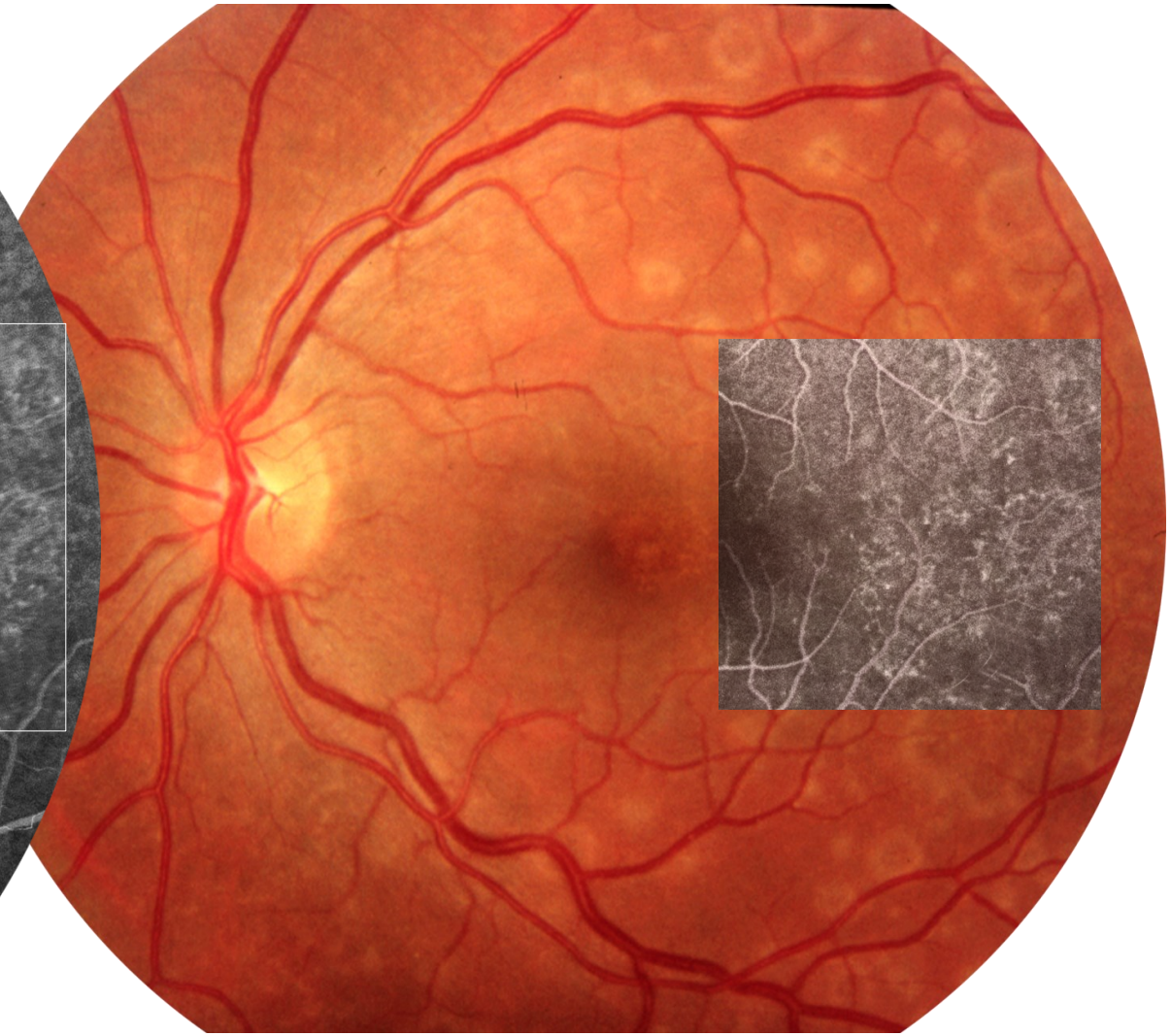


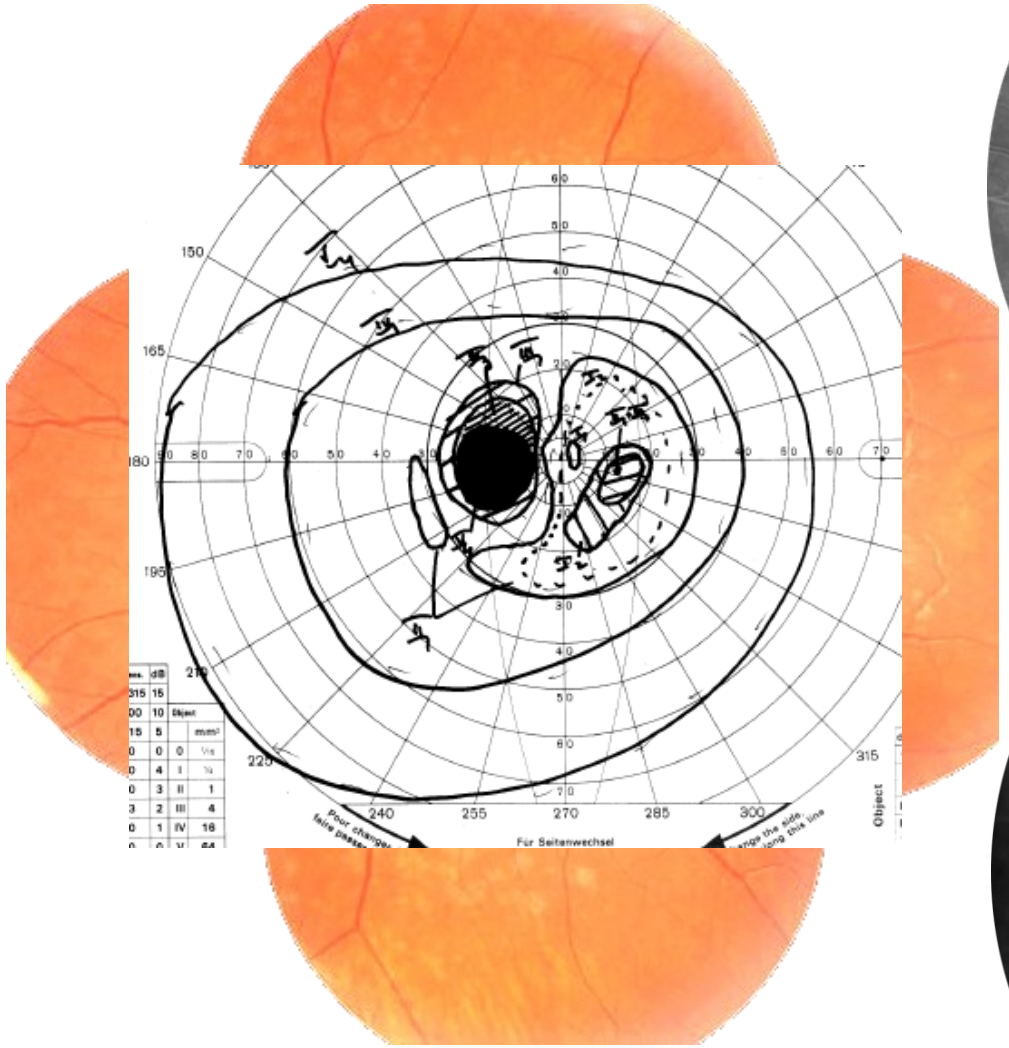
Taches jaunes jusqu'en moyenne périphérie

MEWDS

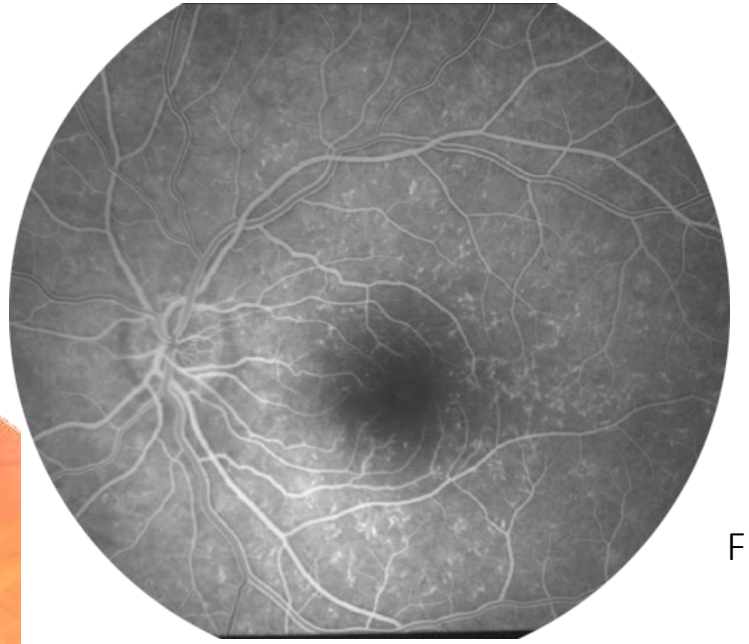
En fluo , "pointillé" en couronne qui dessine le contour des taches



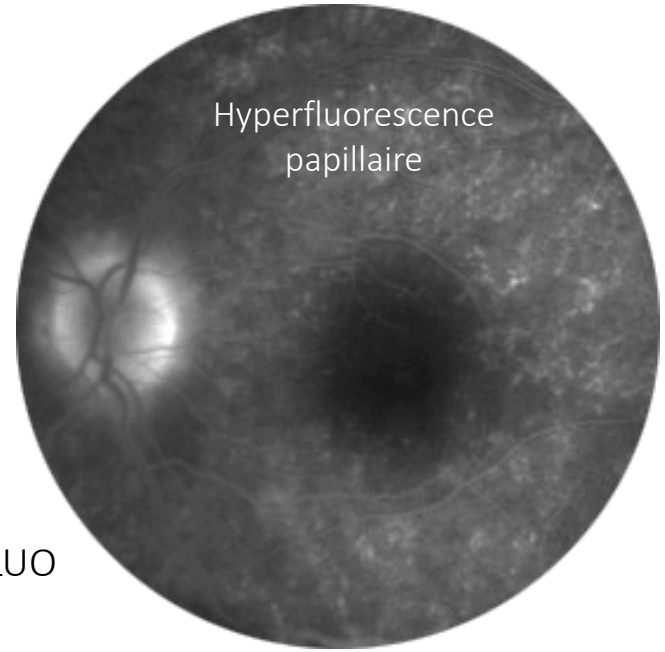




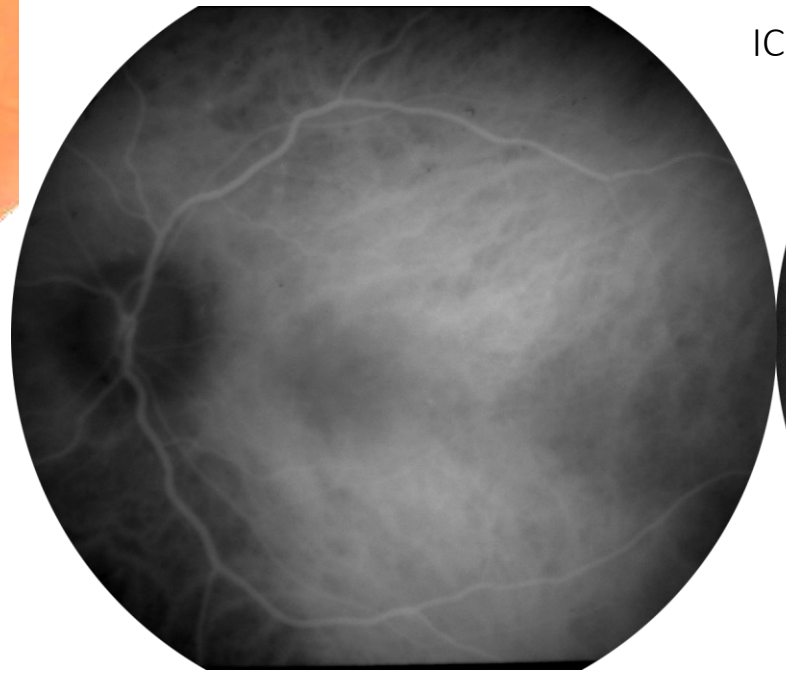
Élargissement de la tache aveugle

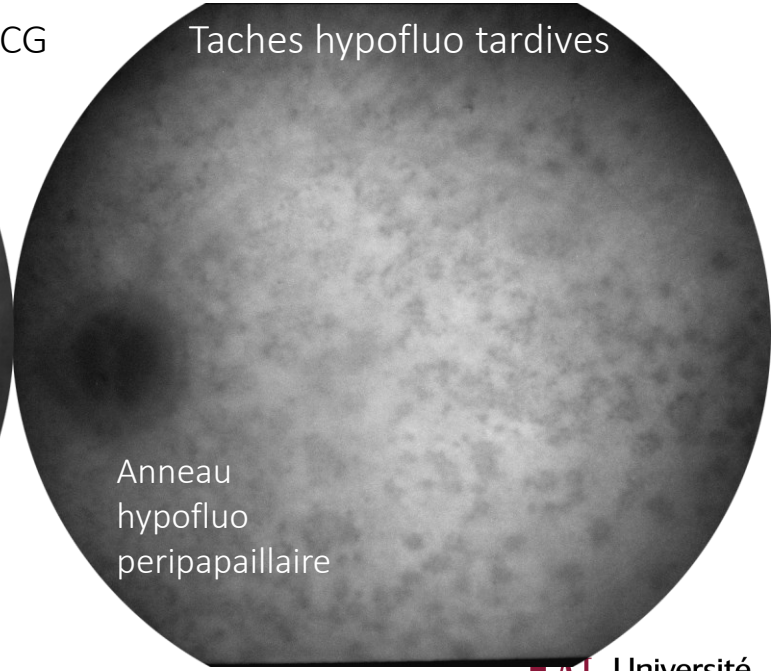


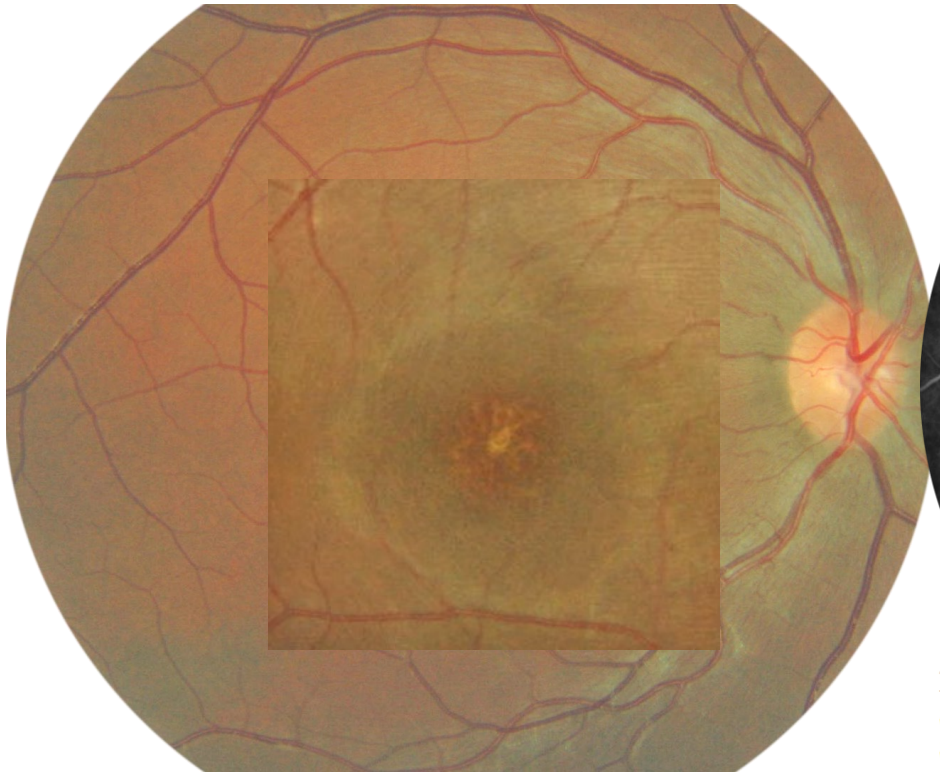
FLUO

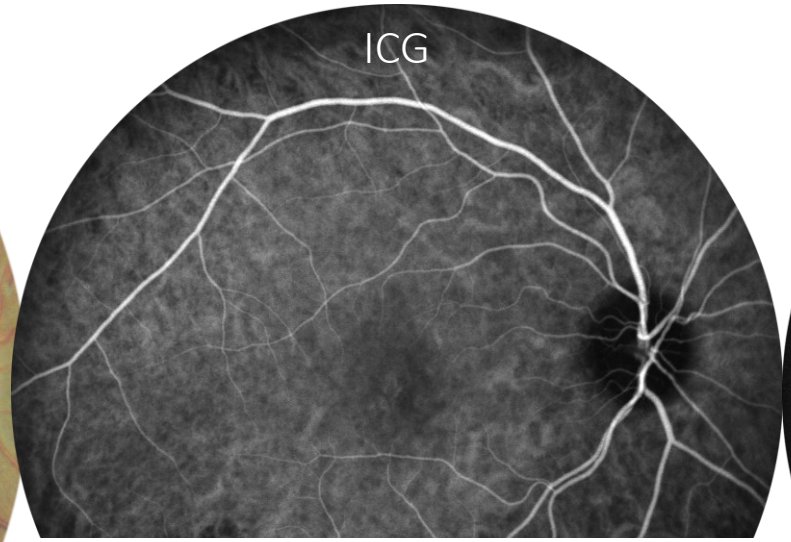


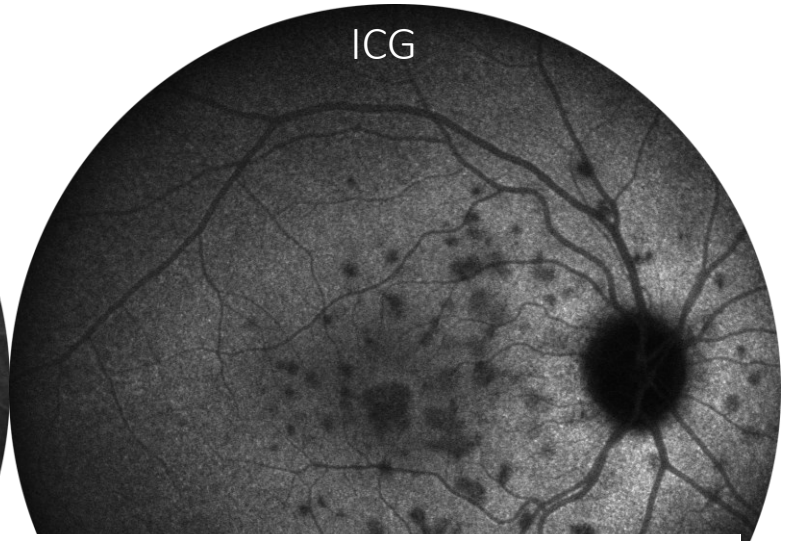
ICG







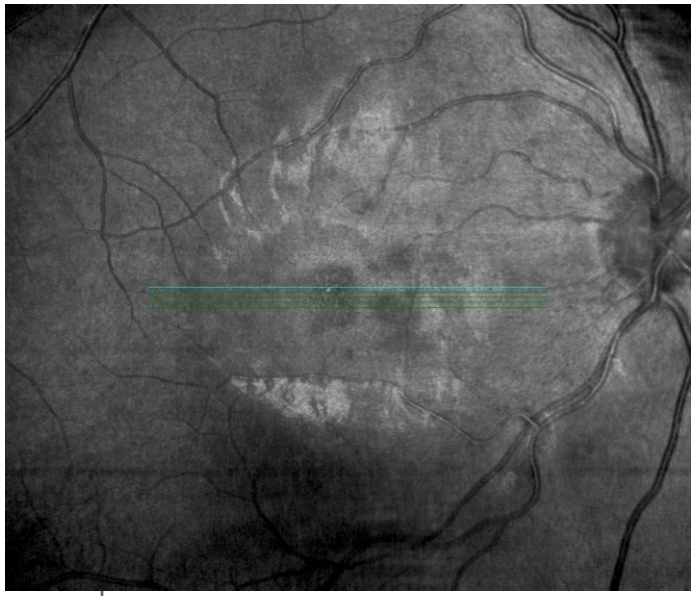


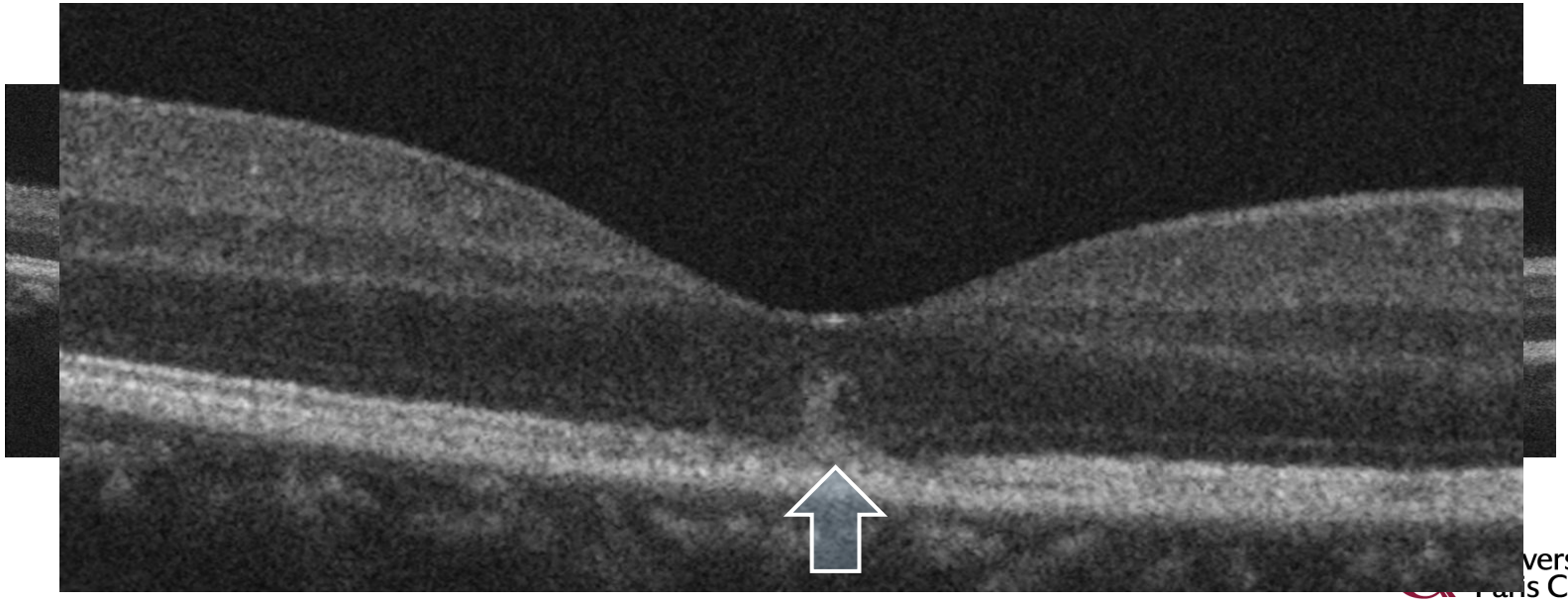


INTRARETINAL HYPERREFLECTIVE LINES

FRANCESCA AMOROSO, MD,* SARAH MREJEN, MD,† ALEXANDRE PEDINIELLI, MD,*
SANDRINE TABARY,† ERIC H. SOUIED, MD, PHD,* ALAIN GAUDRIC, MD,†‡
SALOMON Y. COHEN, MD, PHD*†

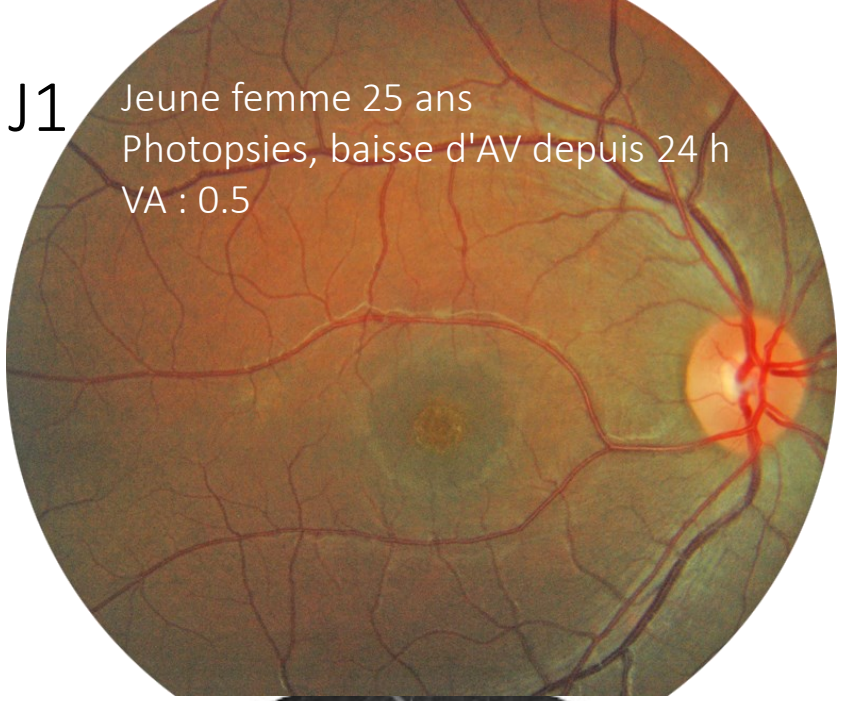
RETINA 2020

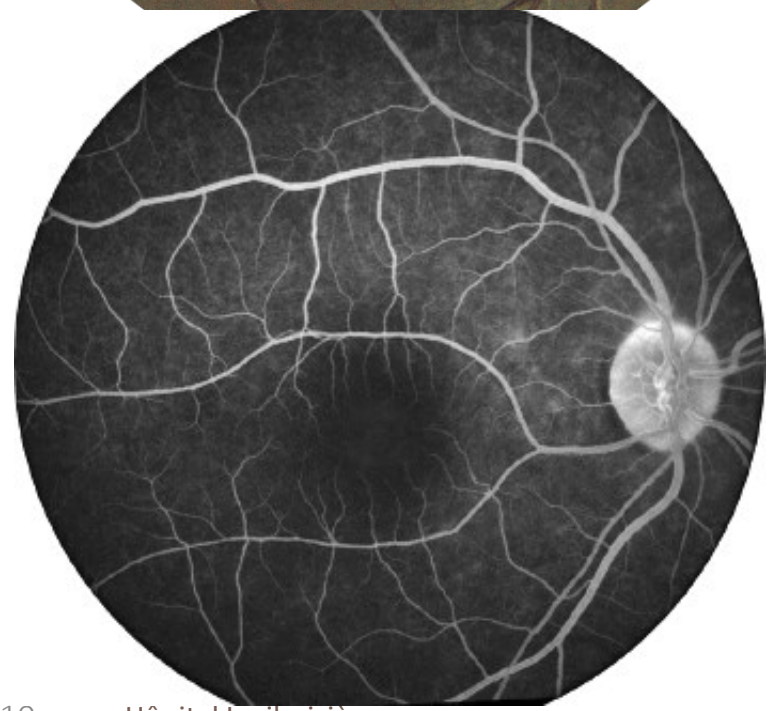




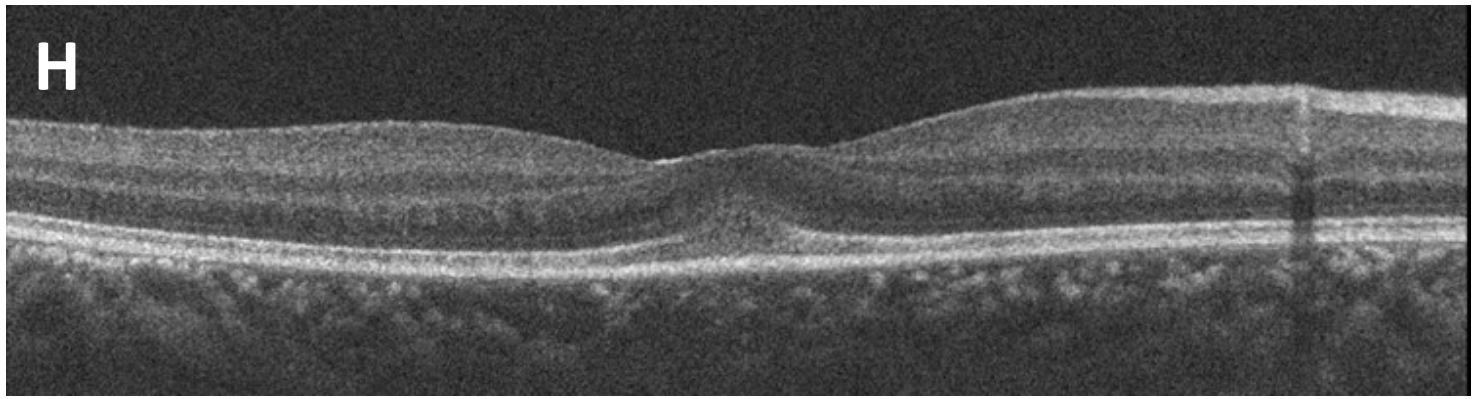
J1

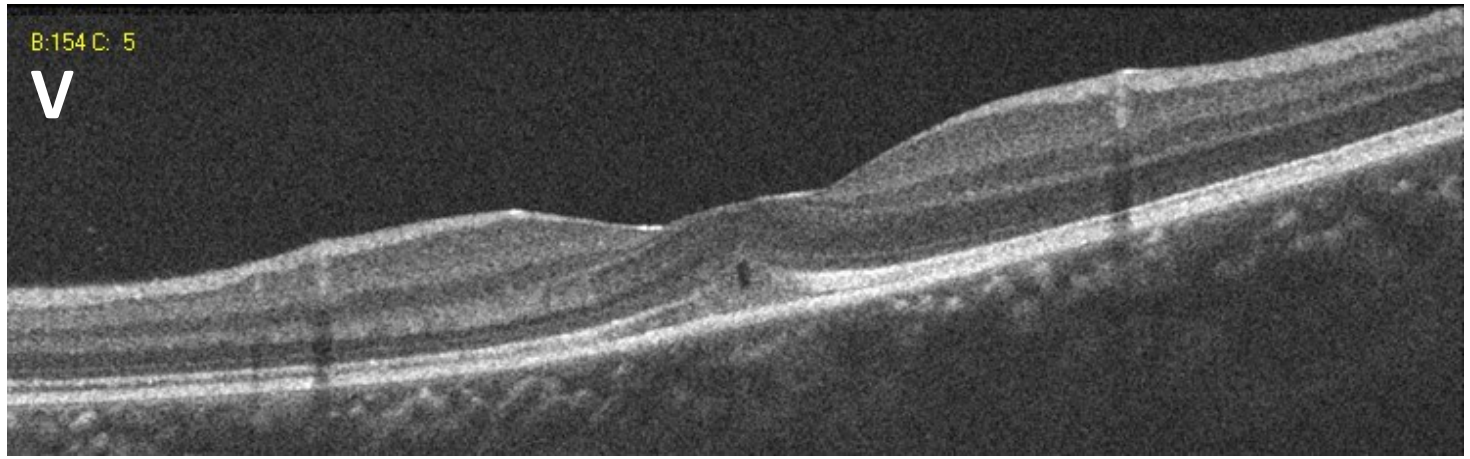
Jeune femme 25 ans
Photopsies, baisse d'AV depuis 24 h
VA : 0.5



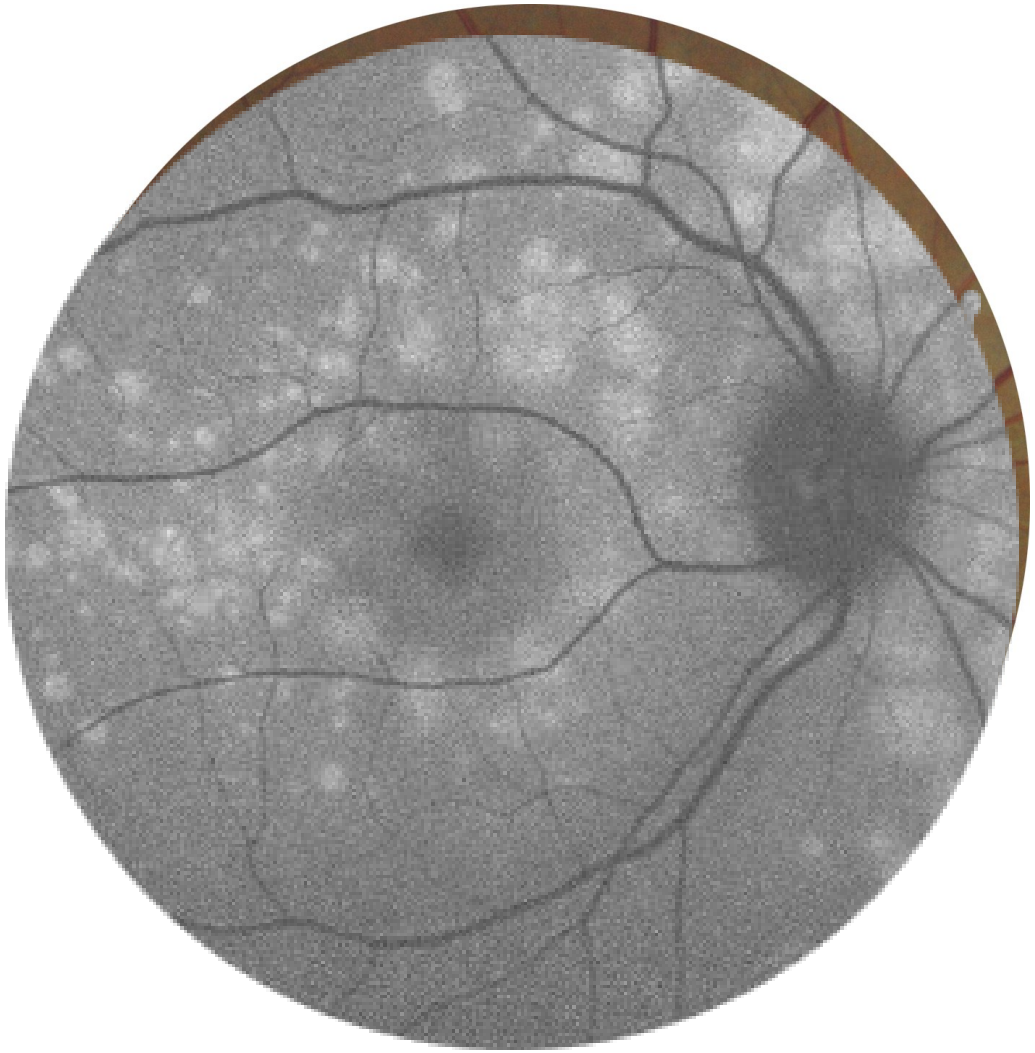


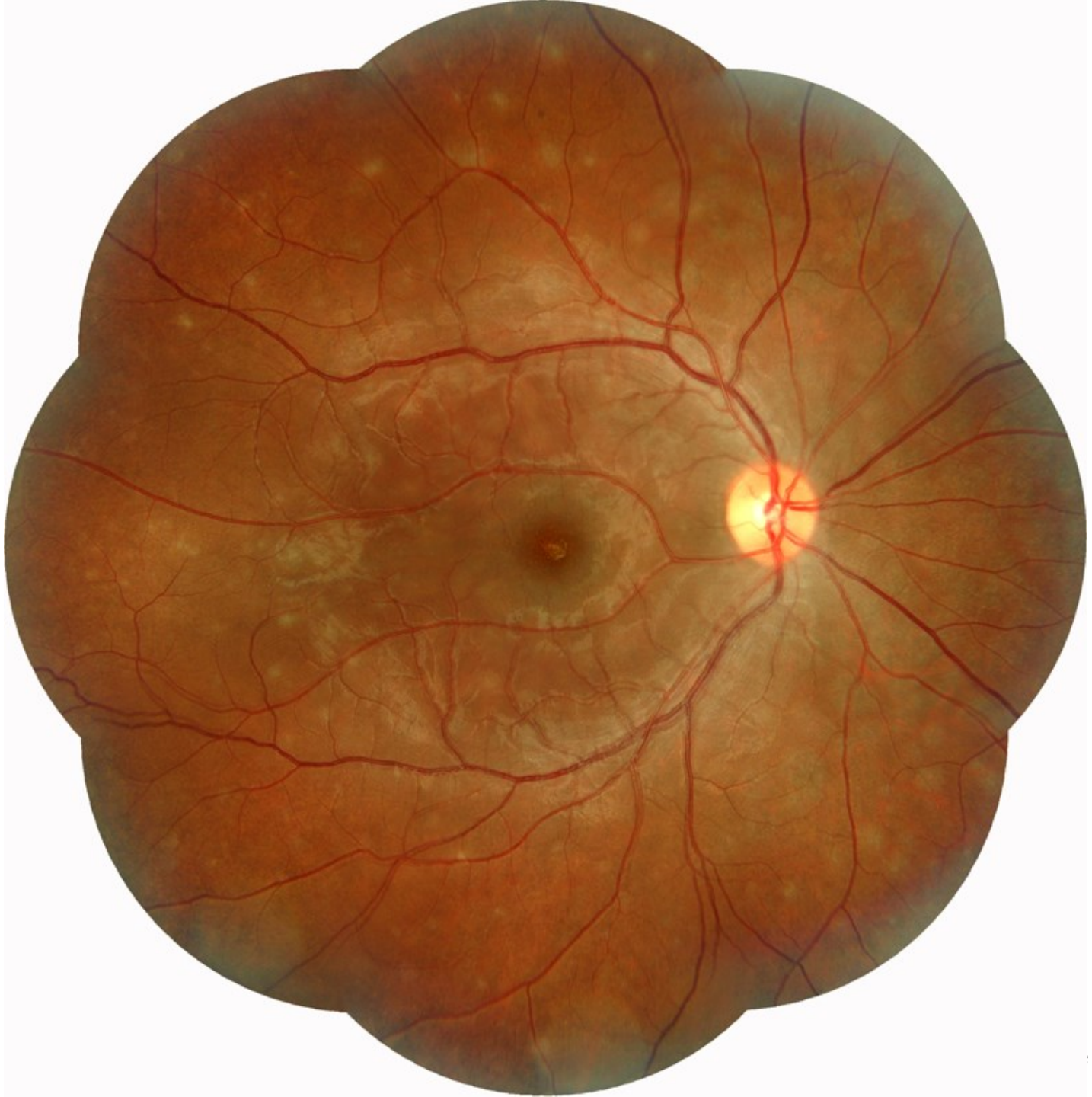
Pas de taches jaunes.
Pas de signes en Angio fluo.
Rupture EZ fovéolaire en OCT
Pas d'ICG pratiquée



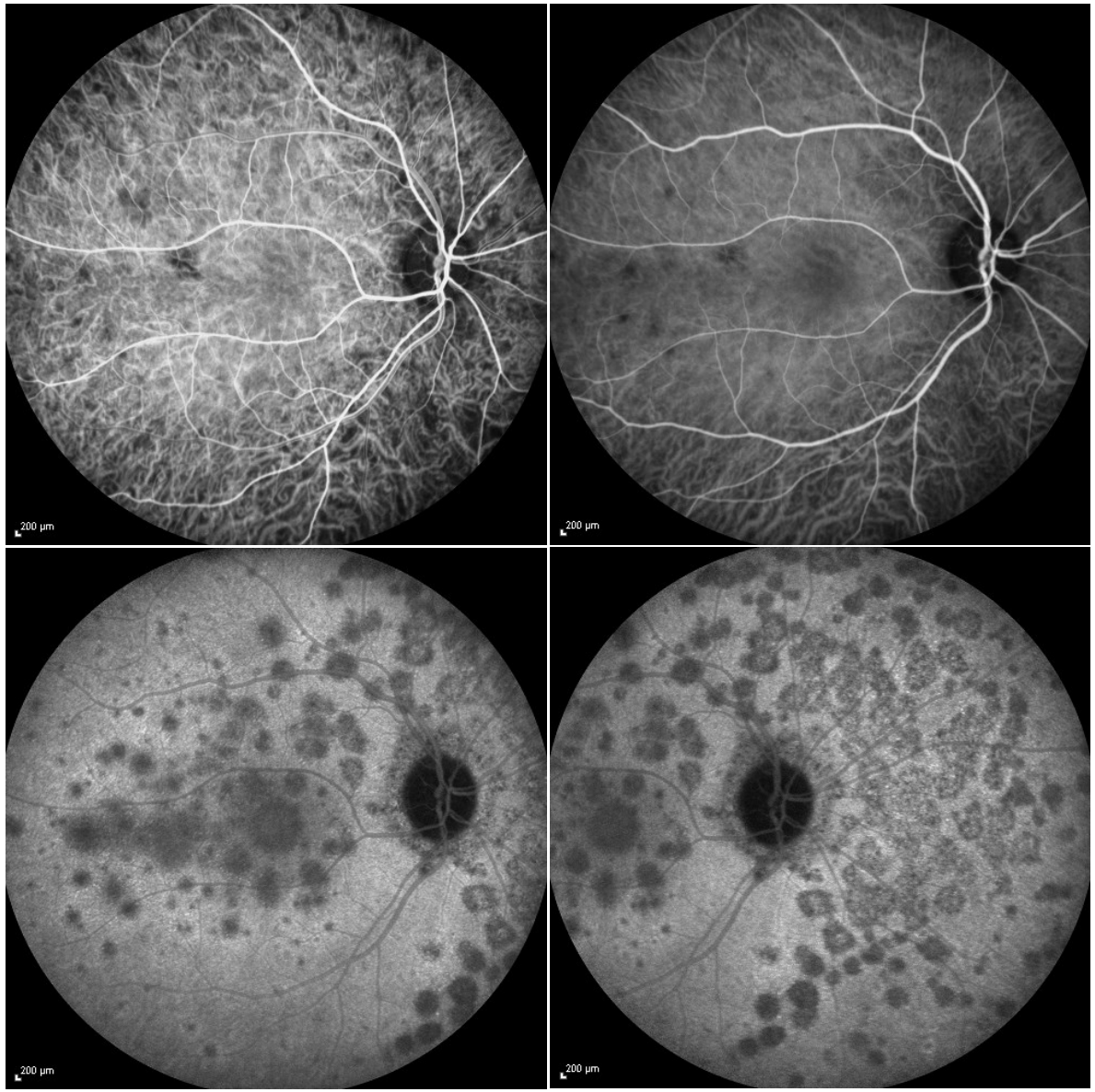


J6 Taches jaunes hyper Autofluo

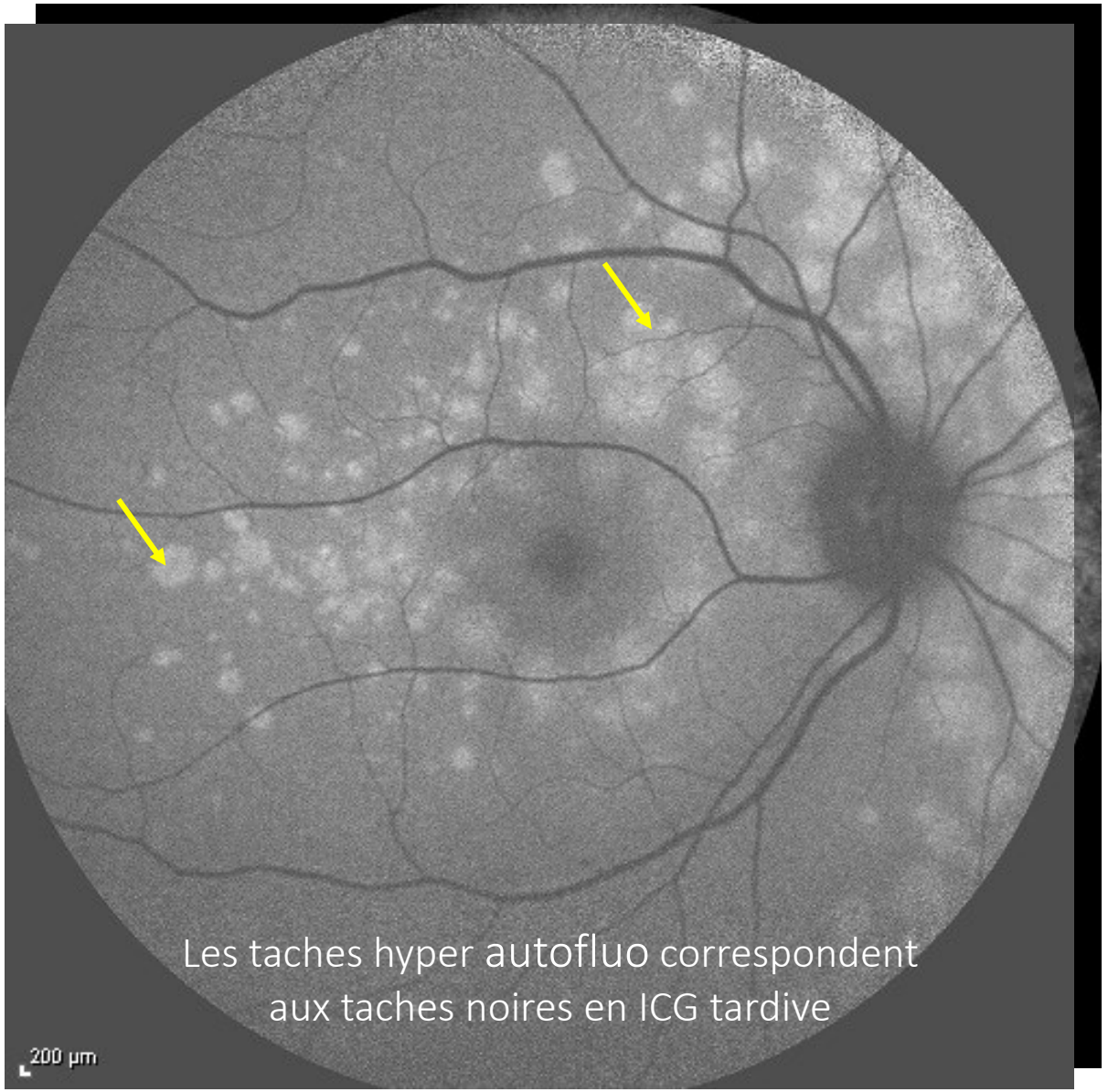




J6


200 µm
200 µm
200 µm
200 µm

ICG vs AF


Les taches hyper autofluo correspondent
aux taches noires en ICG tardive
200 µm

Letters

Freund KB, Mrejen S, Jung J. JAMA Ophtahlmol. Published online Oct 2013.

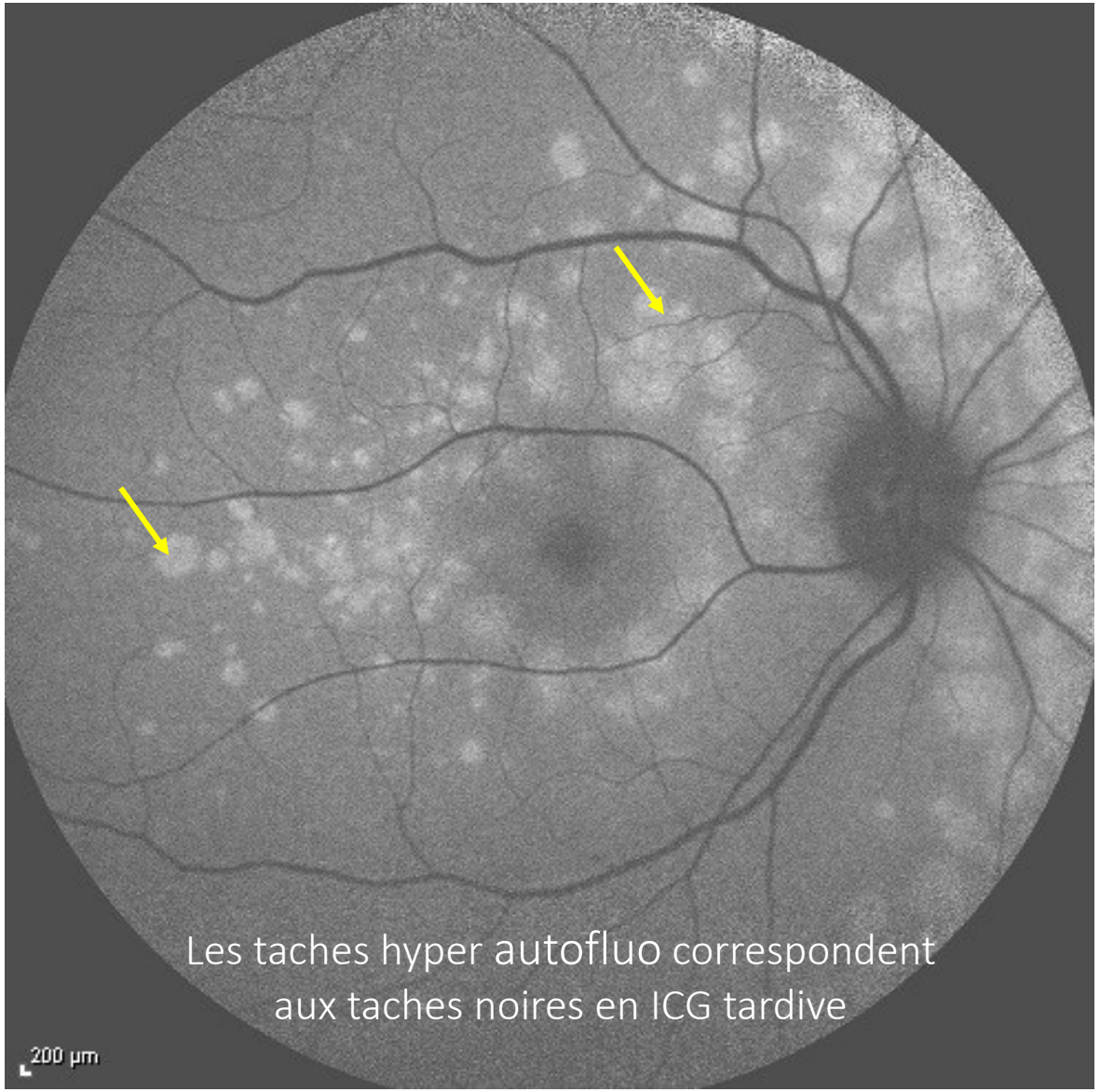
RESEARCH LETTER

Increased Fundus Autofluorescence Related to Outer Retinal Disruption

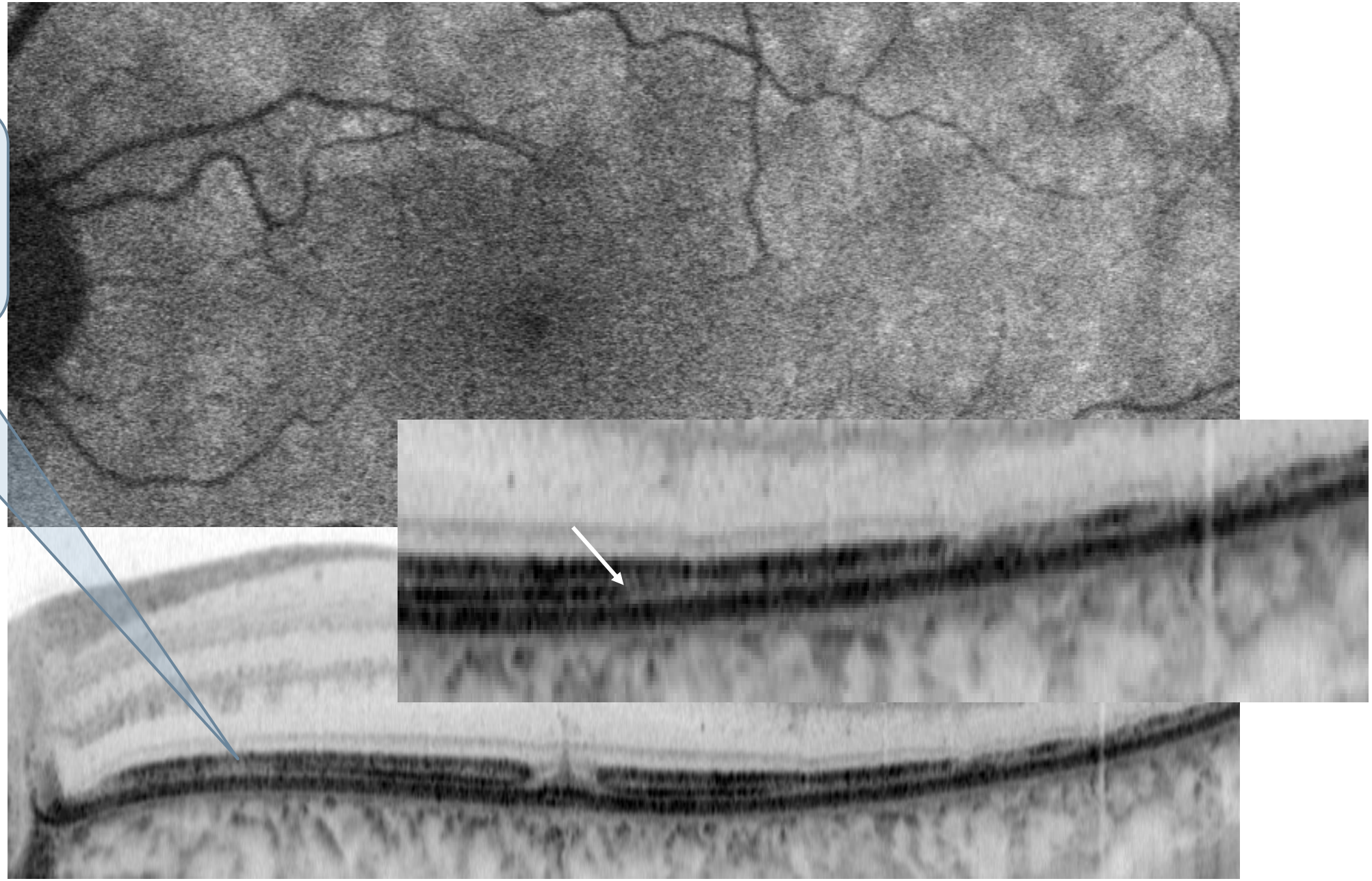
After blue light irradiation, there is photoisomerization of the opsin proteins from the 11-cis to all-trans conformation in the photoreceptor outer segments. This photoisomerization to the all-trans configuration causes a decrease in the optical density of the photopigment in the outer segments of the photoreceptors, resulting in a temporary loss of light absorption properties.

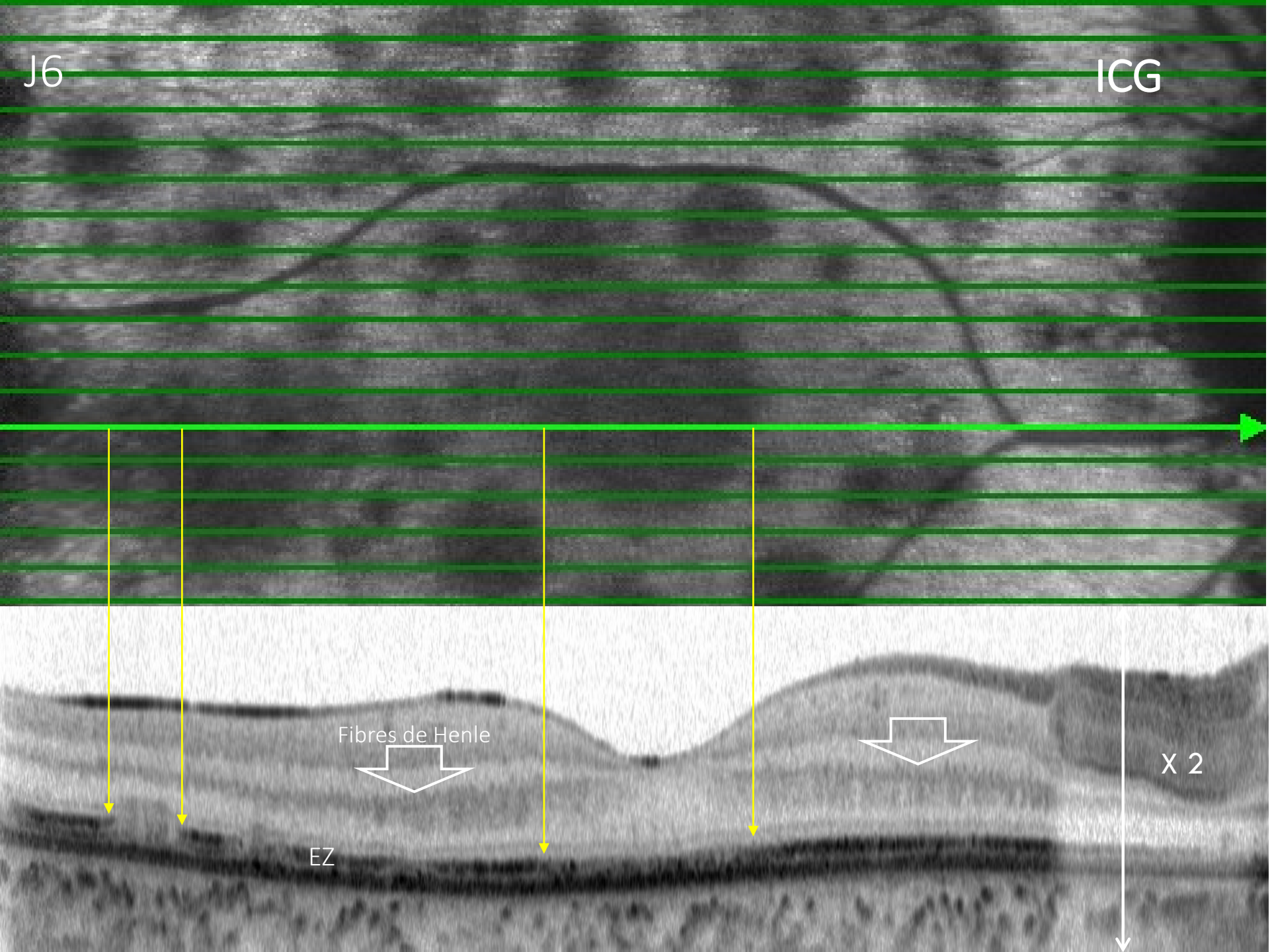
Outer retinal disruption may result in increased autofluorescence due to a window defect as a result of photopigment loss. This mechanism could help explain hyperautofluorescence in a variety of settings including inflammatory entities such as multiple evanescent white dot syndrome and multifocal choroiditis

ICG vs AF



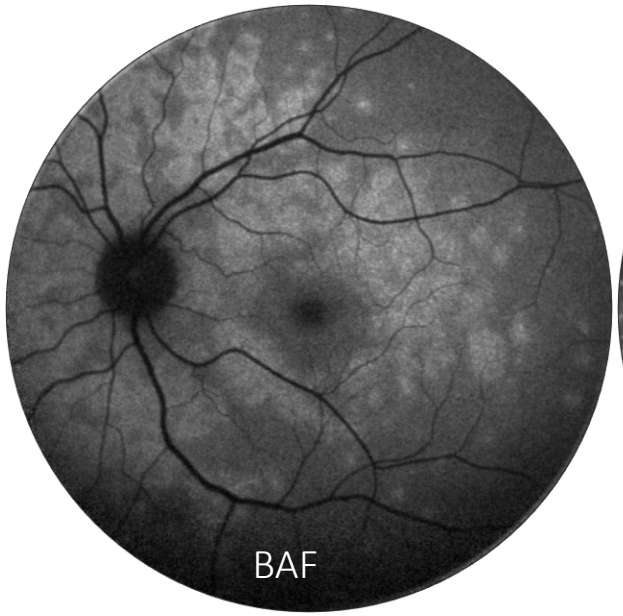
Les taches hyperautofluorescentes correspondent à la disparition des articles externes des photorécepteurs

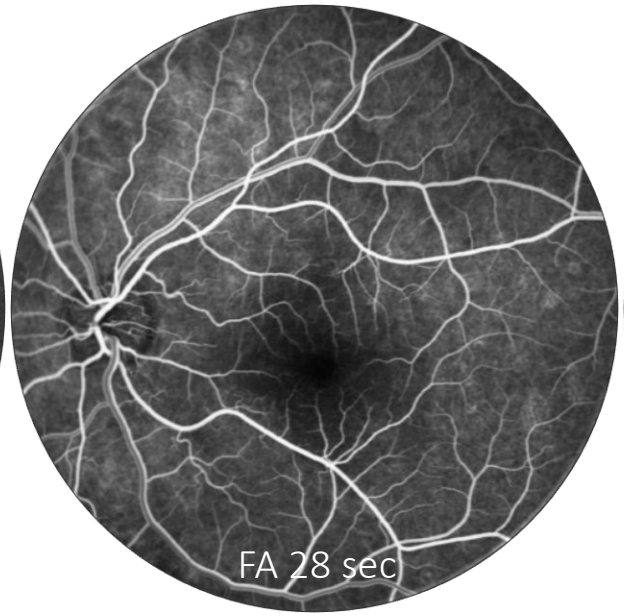


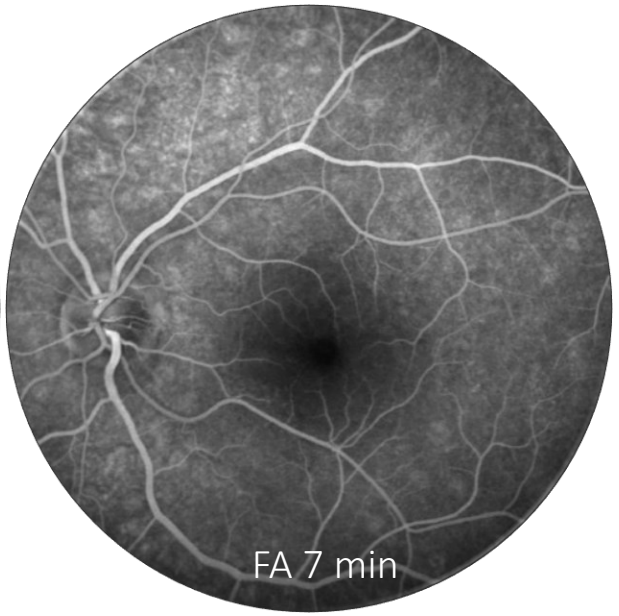


Les taches hypofluorescentes en ICG, correspondent aussi à l'altération des photorécepteurs

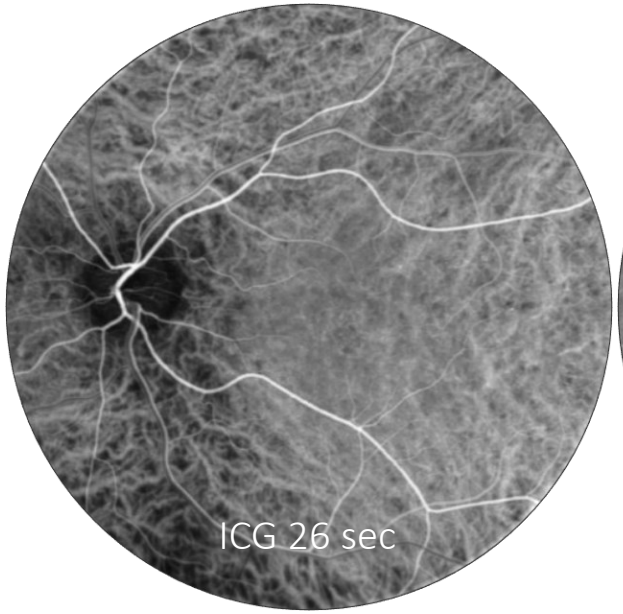
MEWDS

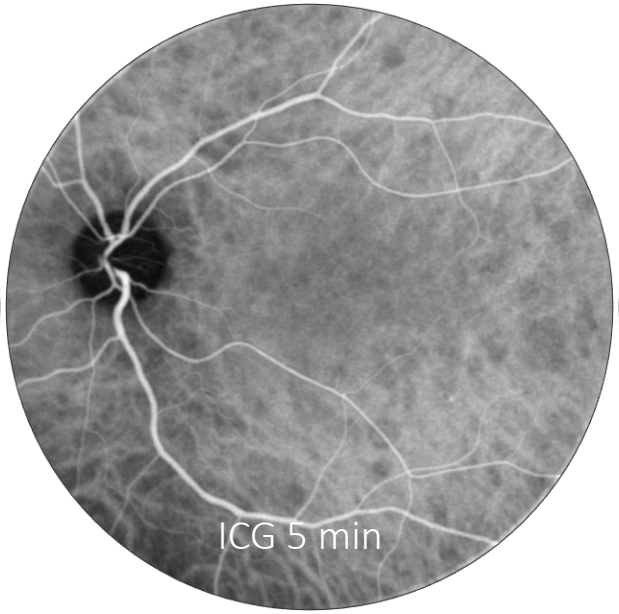


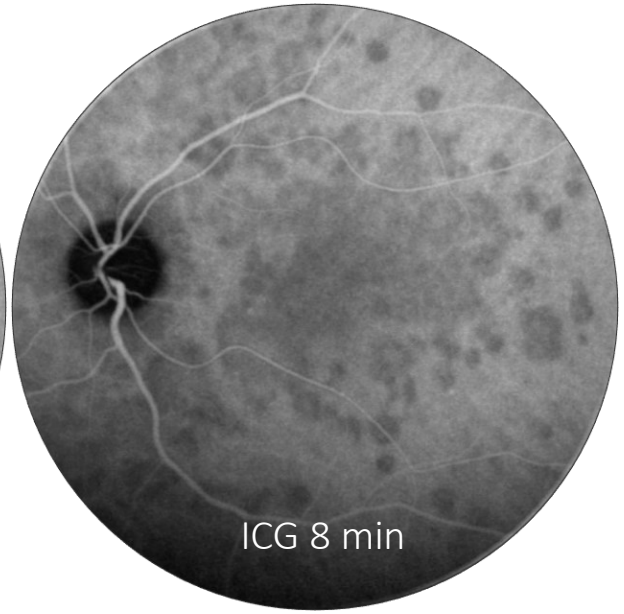


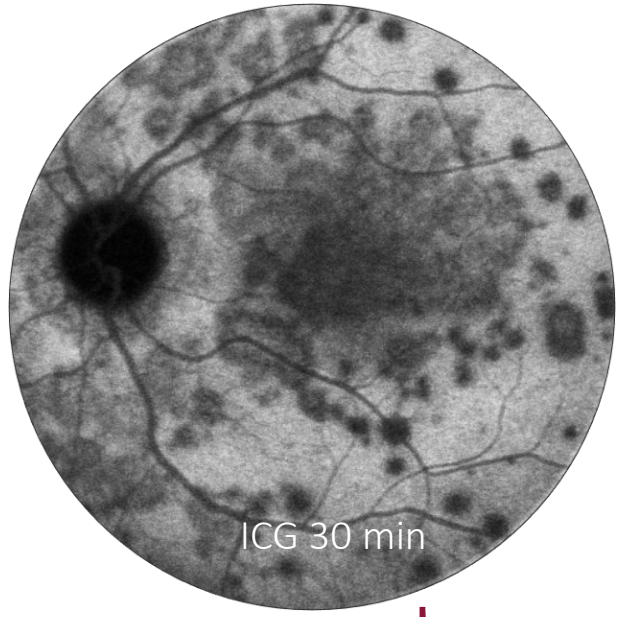


Le temps tardif de l'ICG (30mn) est souvent plus informatif que l'angio Fluo à 10mn

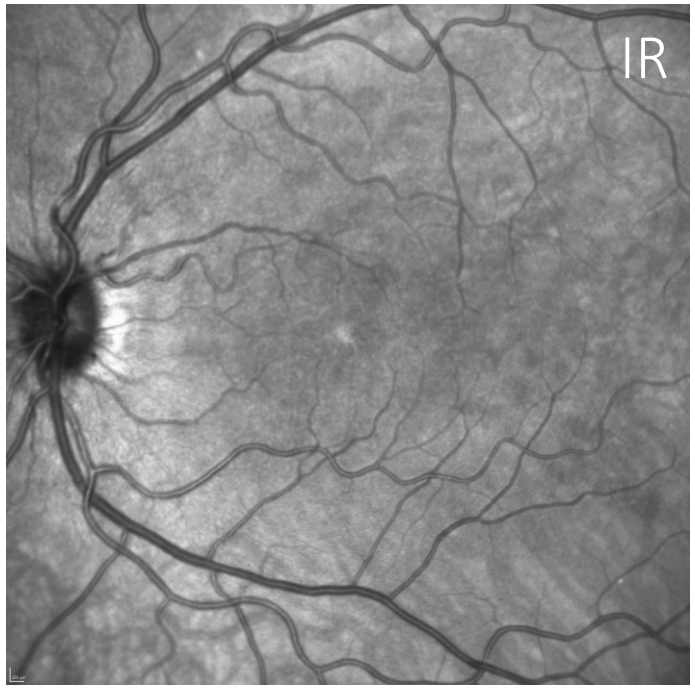


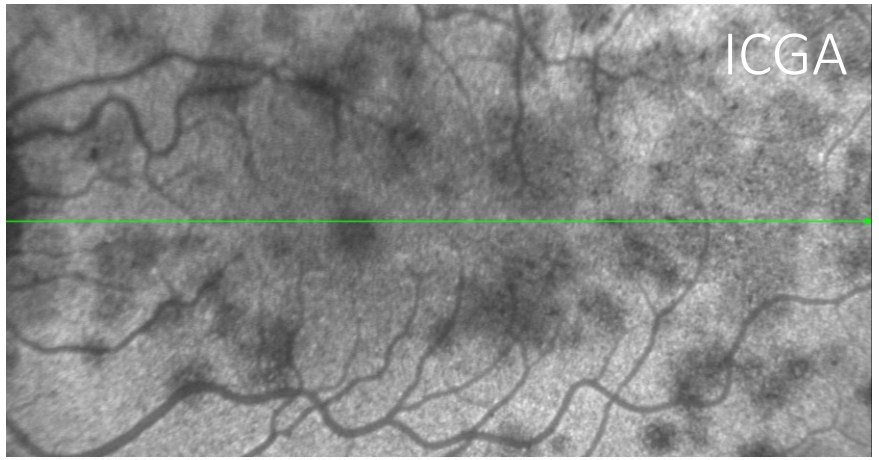


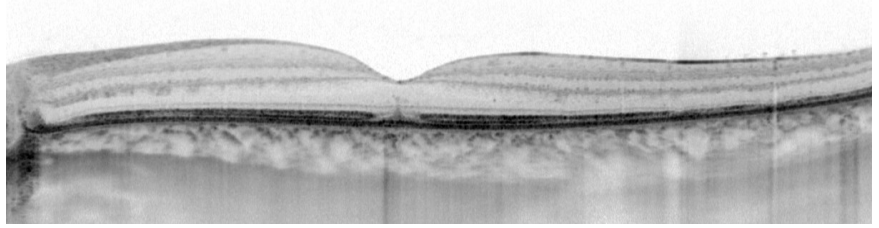


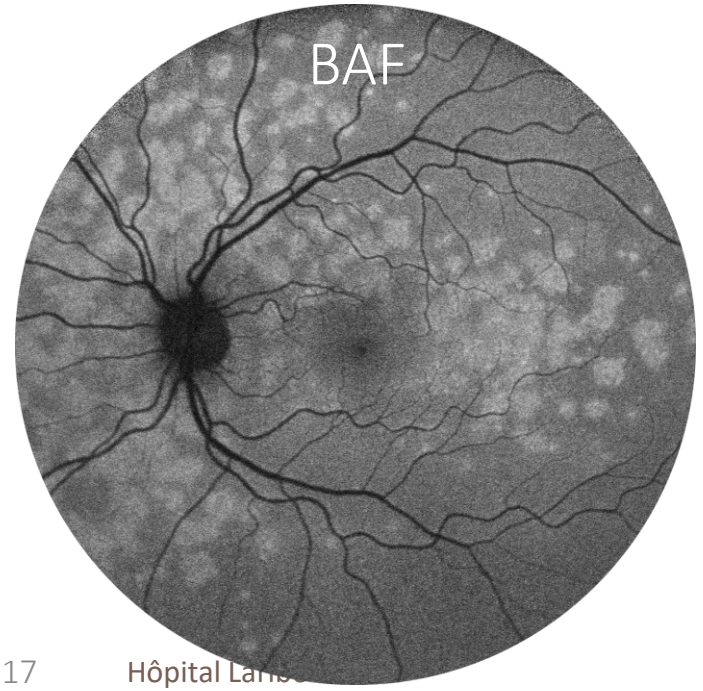


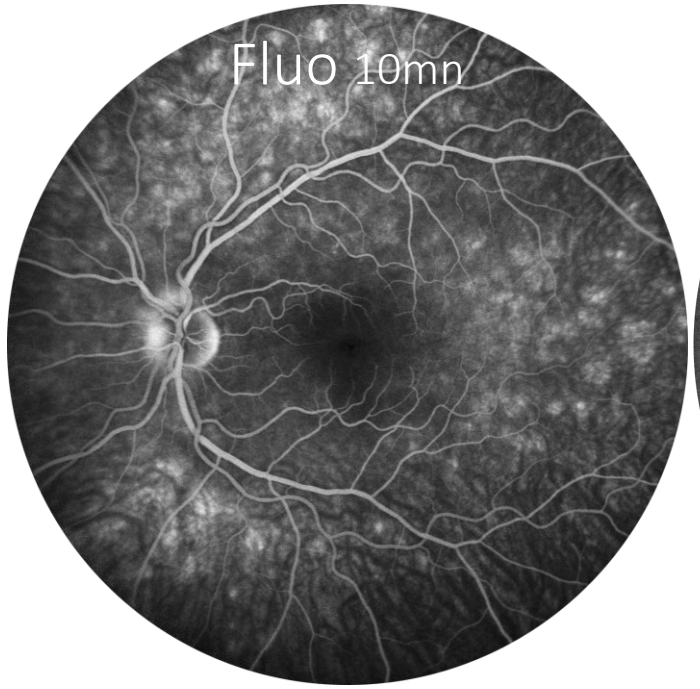
Université Paris Cité

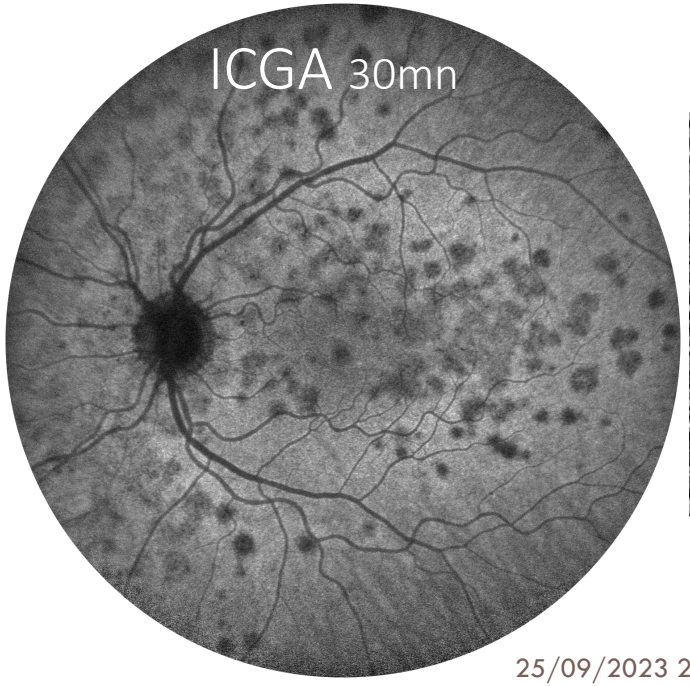












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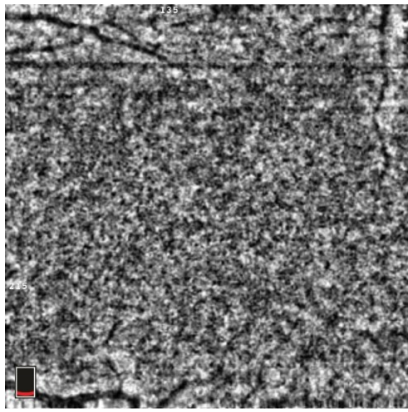
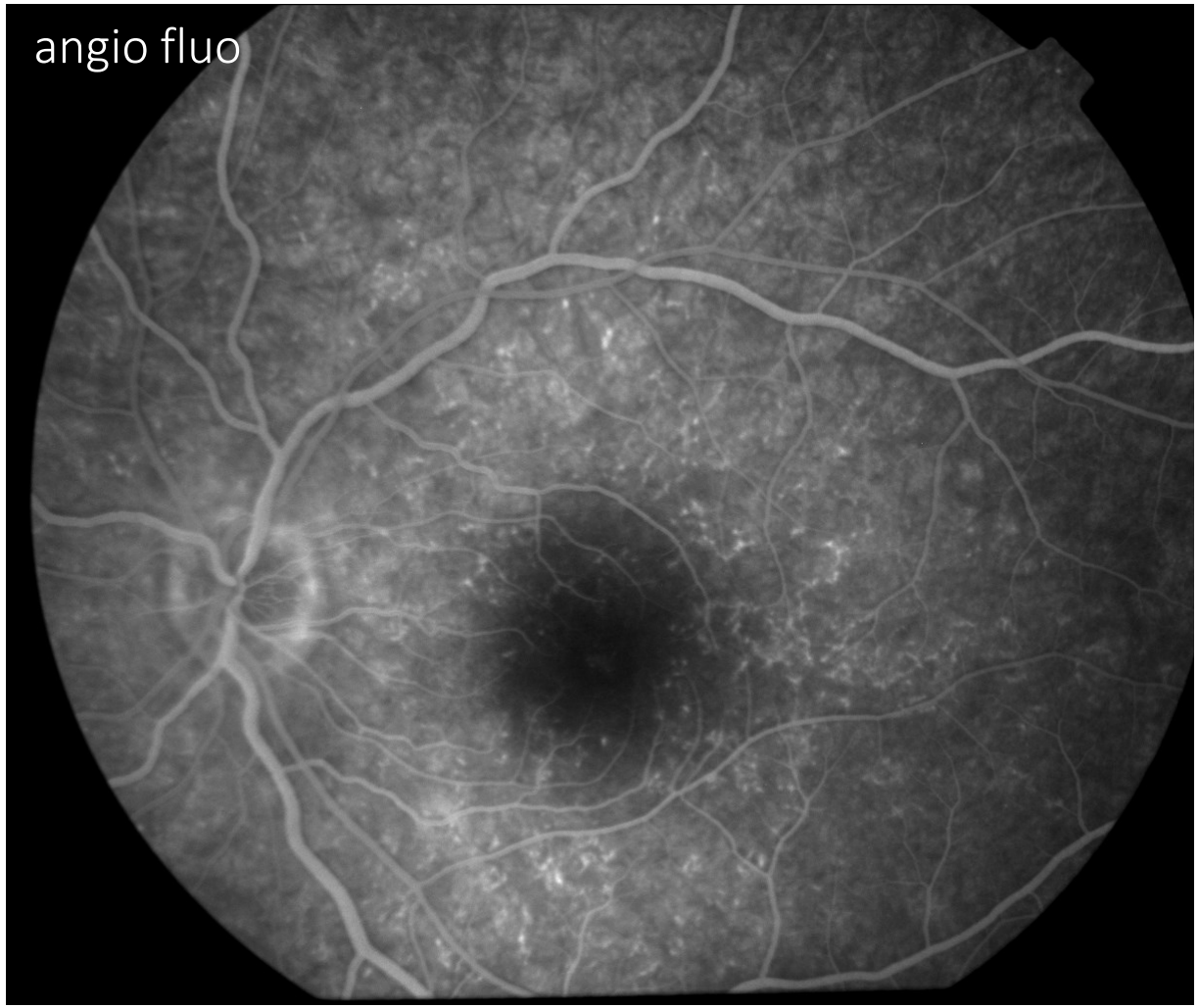
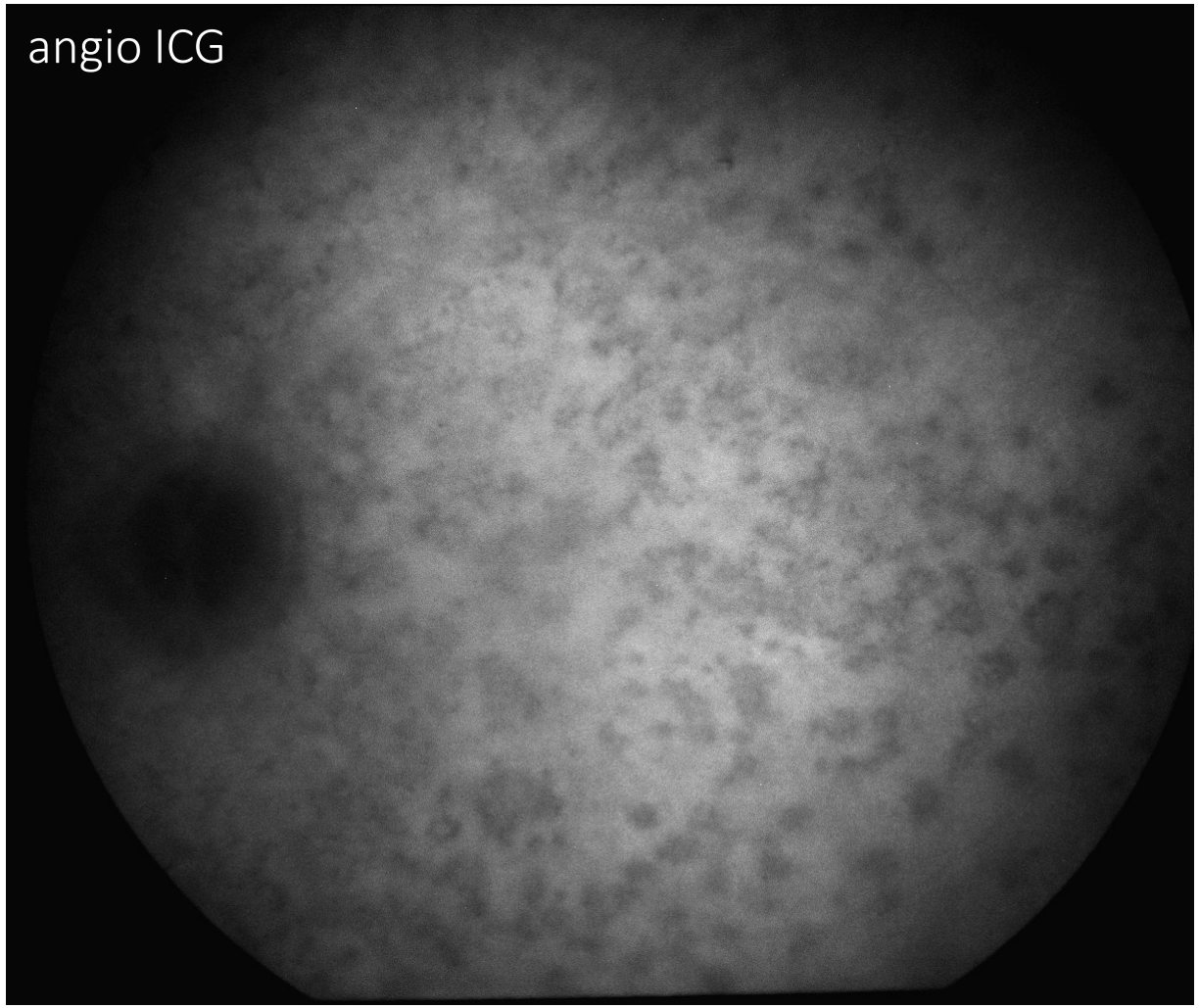


"Dots and Spots"





MEWDS Evolution

- Guérison spontanée
  - En 6 à 8 semaines
  - avec restauration de l'AV
- Des récidives ont été (rarement) décrites
- Dans quelques rares cas , récidive sur le mode d'une Choroidite Multifocale (CMF)

Multiple evanescent white dot syndrome: clinical course and factors influencing visual acuity recovery

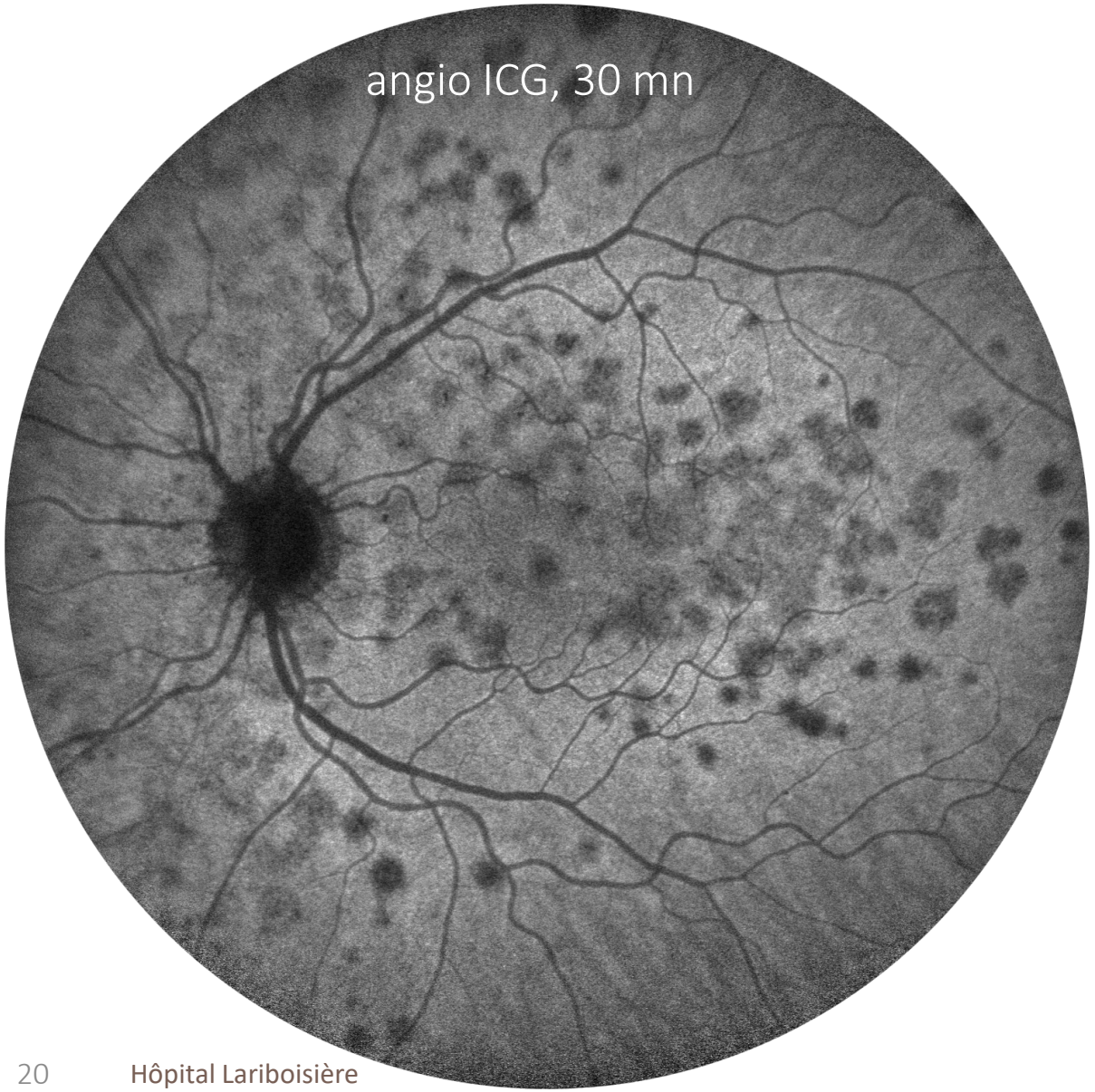
Francesca Bosello [1,2] Mark Westcott,[1] Giuseppe Casalino [1,3] Georgios Agorogiannis,[1] Rocco Micciolo,[4] Angela Rees,[1] Carlos Pavesio[1]

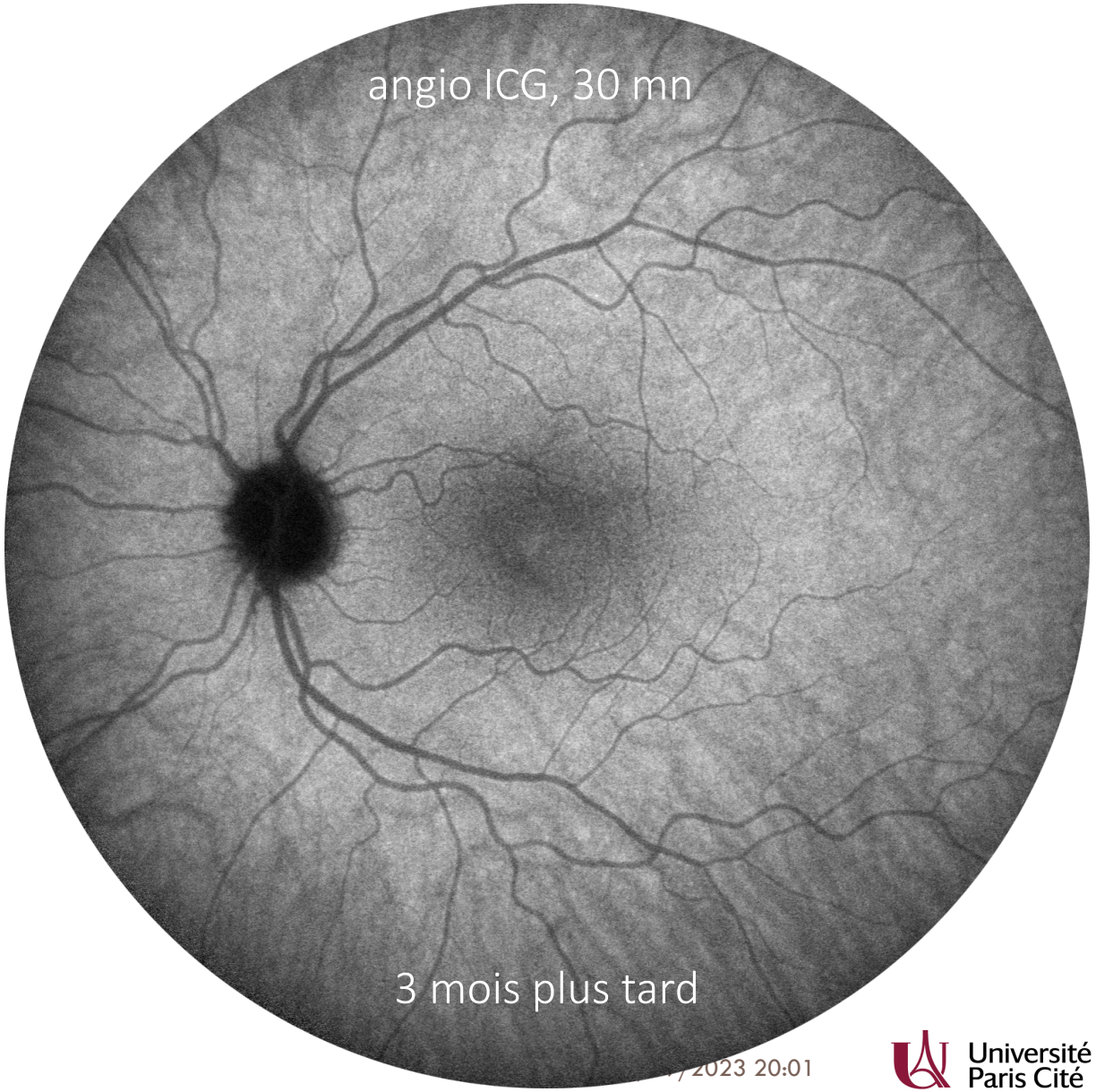
Br J Ophthalmol. 2020

Conclusions Although the majority of cases have a benign prognosis, the clinical spectrum of MEWDS includes incomplete visual recovery. In our series, poor presenting VA and young age were associated with poor VA outcome. Further study is warranted to confirm these findings.

Bien que la majorité des cas aient un pronostic bénin, le spectre clinique du MEWDS comprend une récupération visuelle incomplète. Dans notre série, une mauvaise AV initiale et un jeune âge étaient associés à une mauvaise visdion finale . D'autres études sont justifiées pour confirmer ces observations

MEWDS : evolution





MEWDS associés à des CMF

MULTIPLE EVANESCENT WHITE DOT SYNDROME IN PATIENTS WITH MULTIFOCAL CHOROIDITIS

RICHARD G. BRYAN, MD, PHD, K. BAILEY FREUND, MD, LAWRENCE A. YANNUZZI, MD, RICHARD F. SPAIDE, MD, SHEAU J. HUANG, MD, DANIELLE L. COSTA, MD

RETINA 22:317–322, 2002

Simultaneous Appearance of Multiple Evanescent White Dot Syndrome and Multifocal Choroiditis Indicate a Common Causal Relationship

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Ocular Immunology & Inflammation, 17(5), 325–327, 2009

Expanding the Clinical Spectrum of Multiple Evanescent White Dot Syndrome with Overlapping Multifocal Choroiditis

Hyun Goo Kang, MD[a,b], Tae Young Kim MD[a], Min Kim, MD, PhD[a,b], Suk Ho Byeon, MD, PhD[a,b], Sung Soo Kim, MD, PhD[a,b], Hyoung Jun Koh, MD, PhD[a,b], Sung Chul Lee, MD, PhD[b], and Christopher Seungkyu Lee, MD, PhD[a,b]

Ocul Immunol Inflamm. Published online 2020:1-9.

MEWDS "secondaires"

A Multiple Evanescent White Dot Syndrome–like Reaction to Concurrent Retinal Insults

Maria Vittoria Cicinelli, MD,[1,2,3,*] Omar Mohamed Hassan, MD,[1*] Manjot K. Gill, MD,[1] Debra Goldstein, MD,[1] Maurizio Battaglia Parodi, MD,[2,3] Lee M. Jampol, MD[1]

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RETINA 42:227–235, 2022

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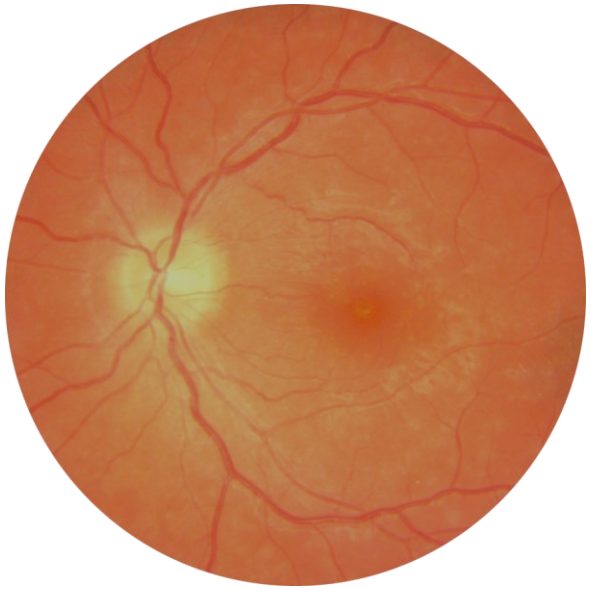
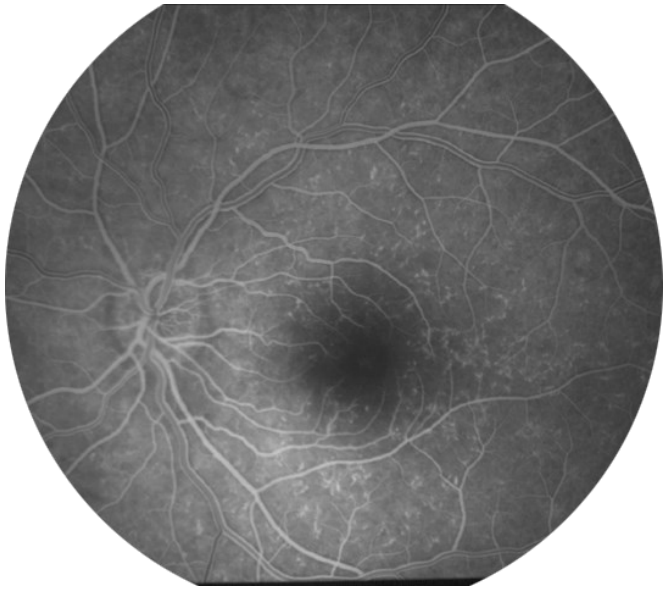
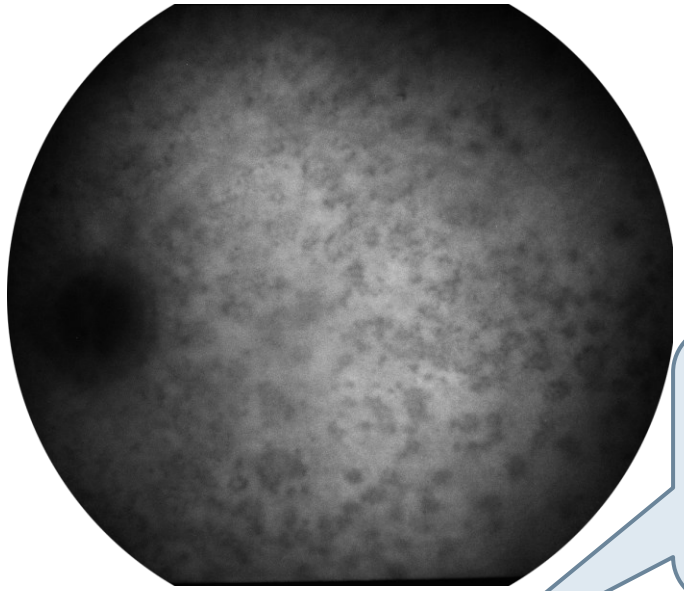
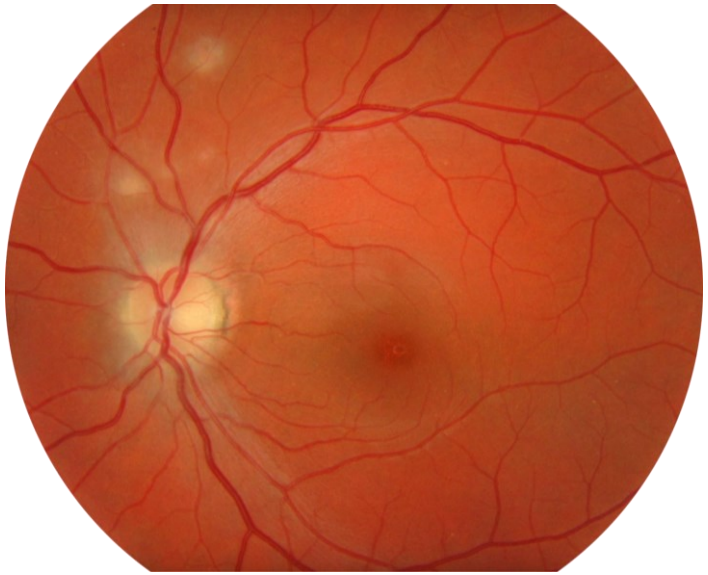
CMF survenant après MEWDS

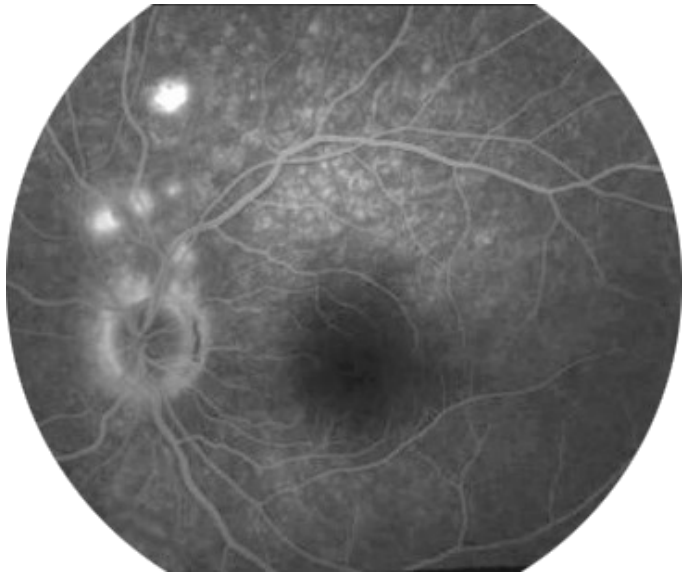


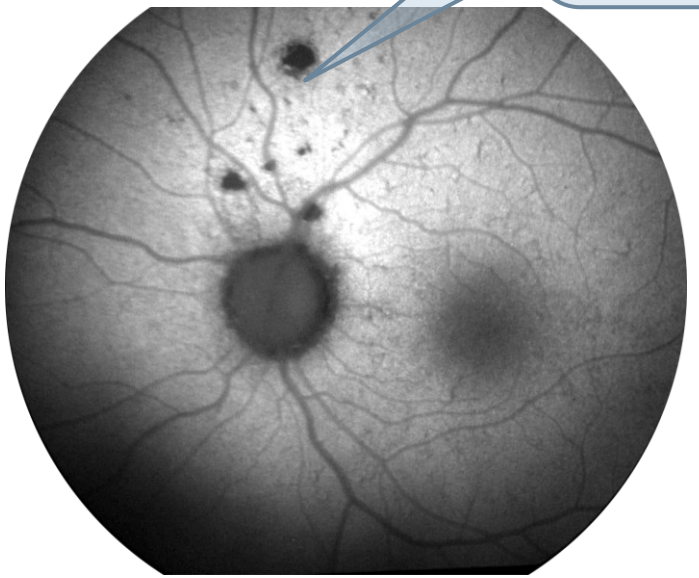




MEWDS typique

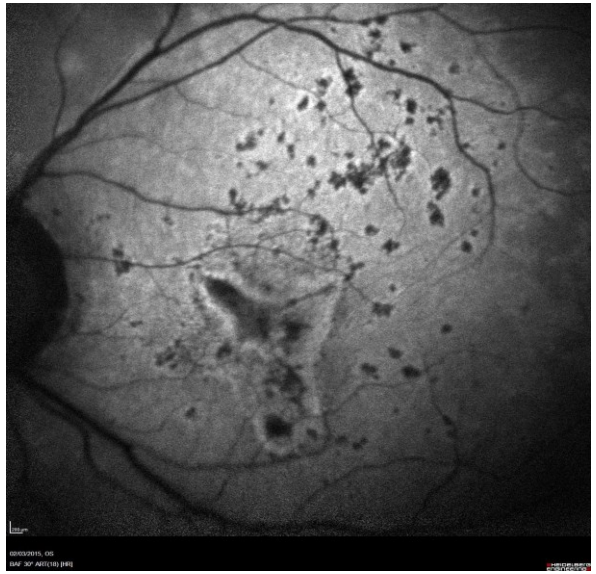




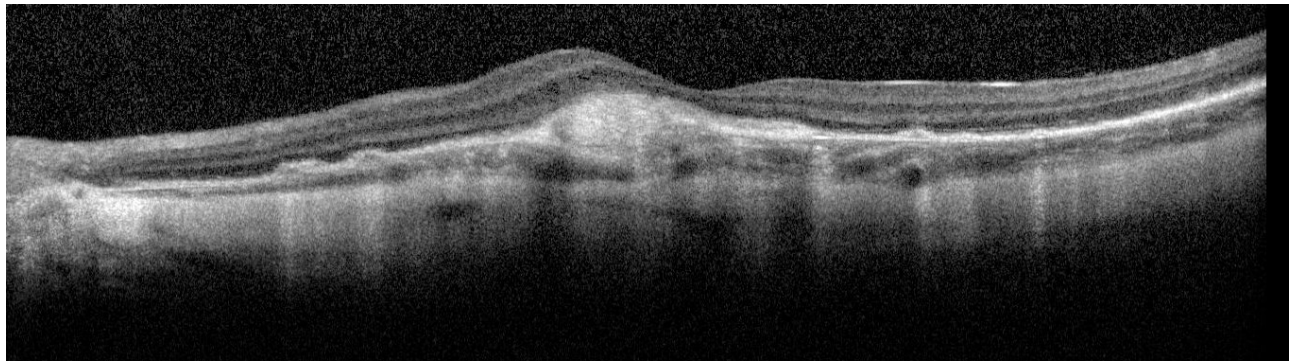


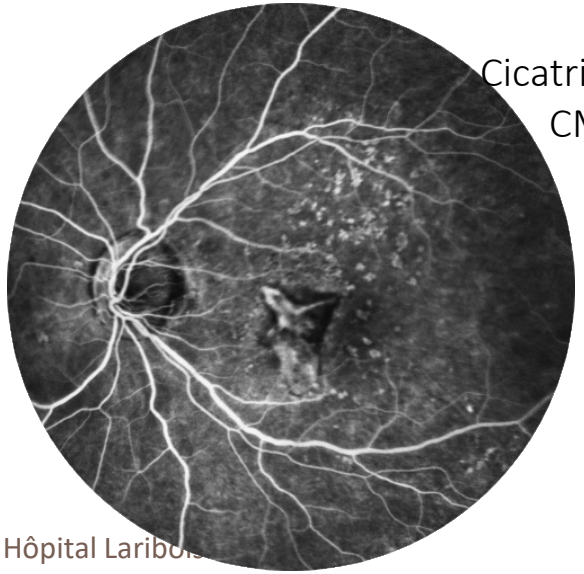
3 ans plus tard , choroidite multifocale + quelques taches hypofluorescentes en ICG

MEWDS sur antécédents de CMF

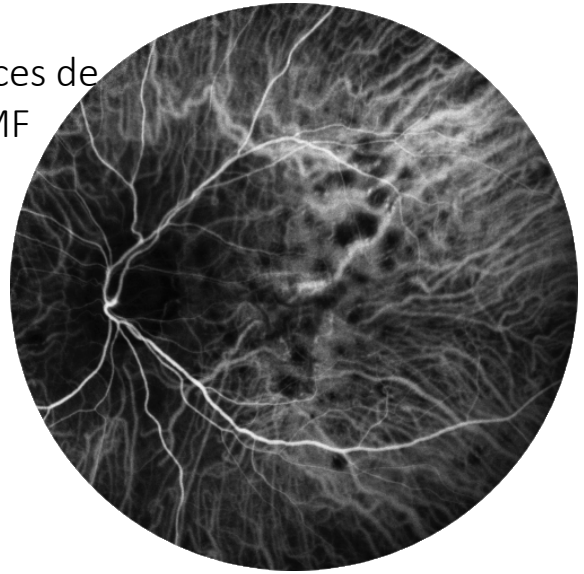


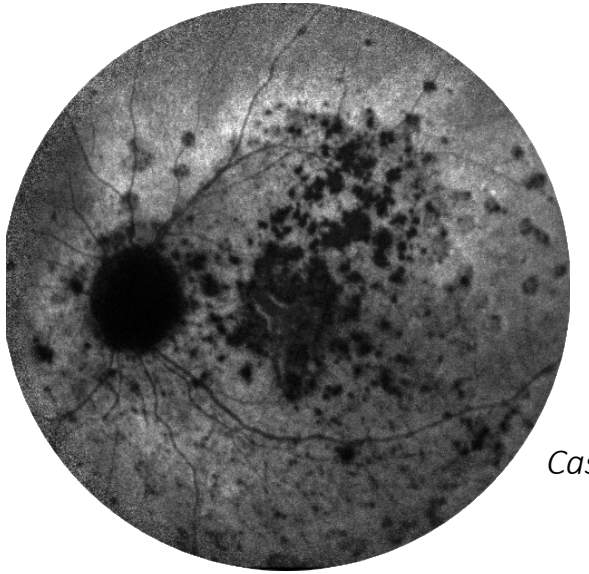
- Cicatrice de CMF avec NVC fibrosés
  - 1 an plus tard : baisse d'AV : MEWDS !





Cicatrices de CMF





1 an plus tard
ICGA 30 mn

Cas du Dr MH Errera XV-XX, Pittsburg

MEWDS : associations

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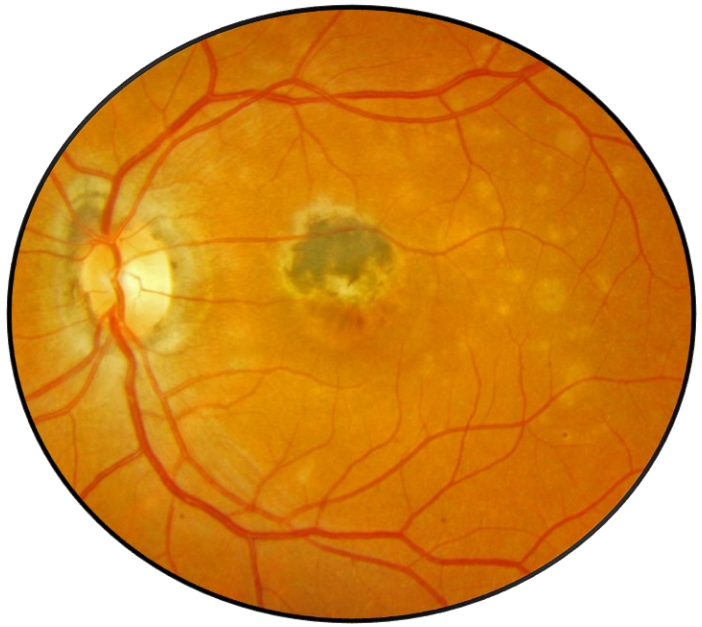

MEWDS "secondaires"

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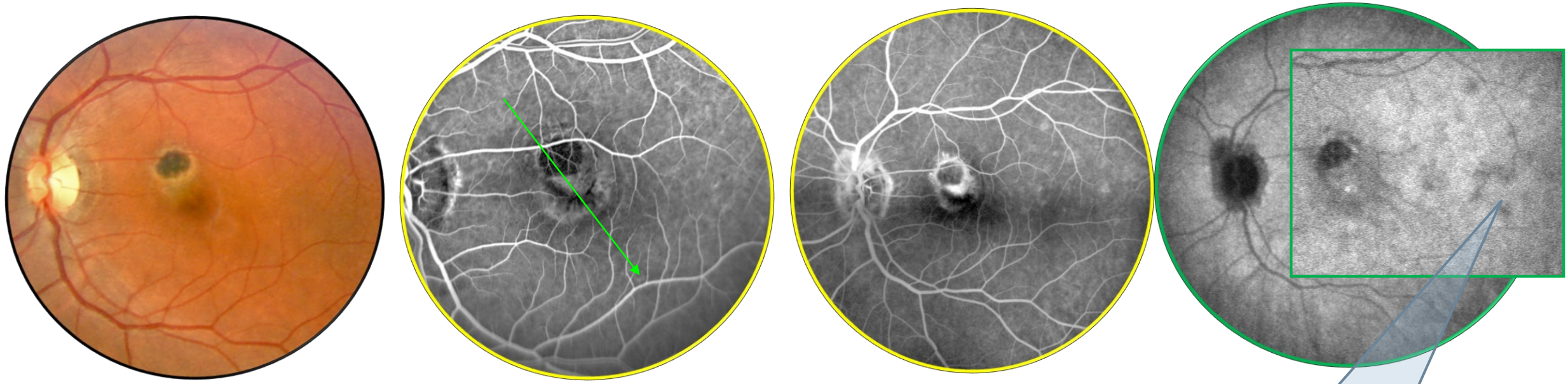
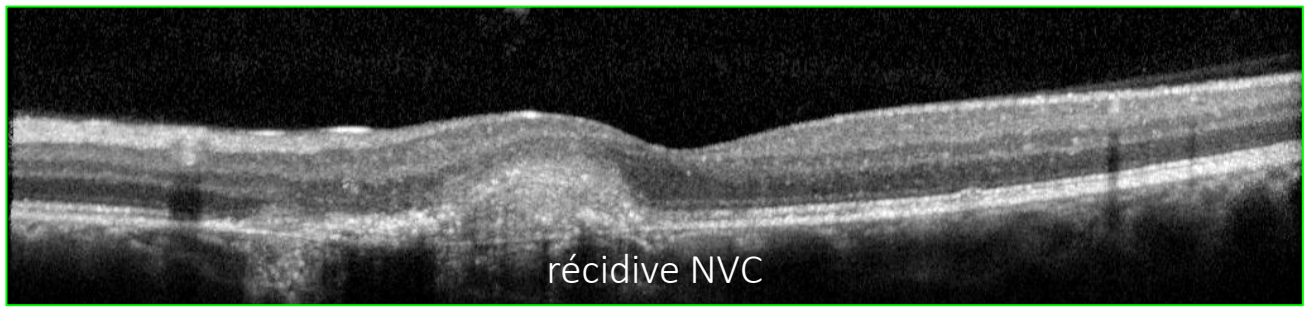
MEWDS secondaire à des NVC

- Apparition de NVC sur cicatrice de laser pour NVC idiopathiques

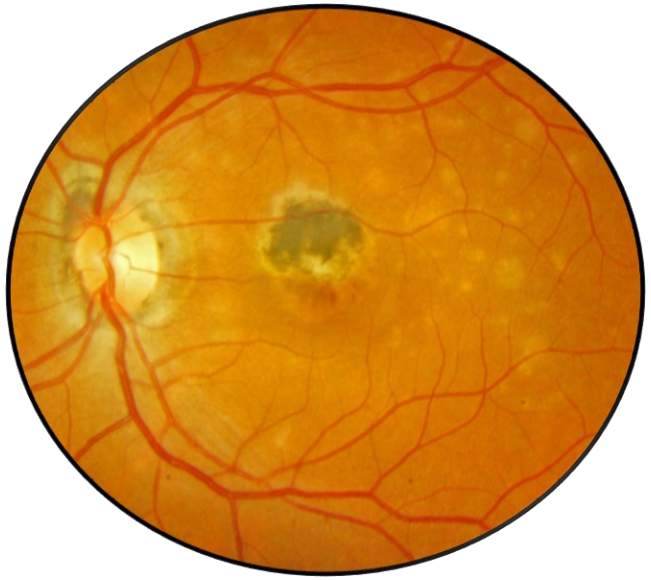


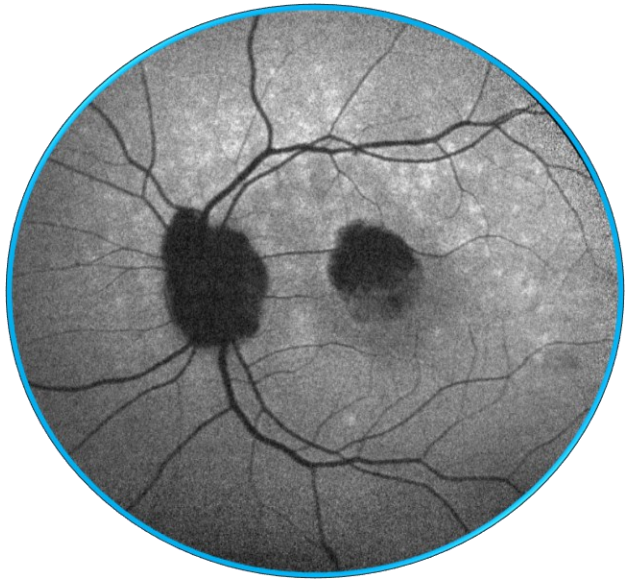


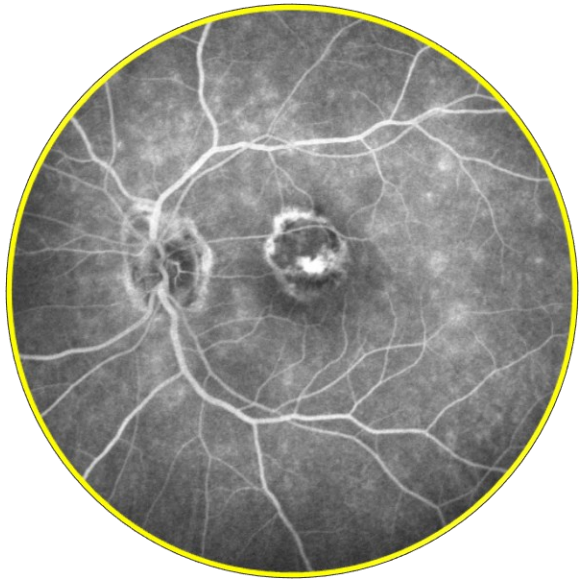
Apparition de discrètes taches hypofluorescentes en ICG

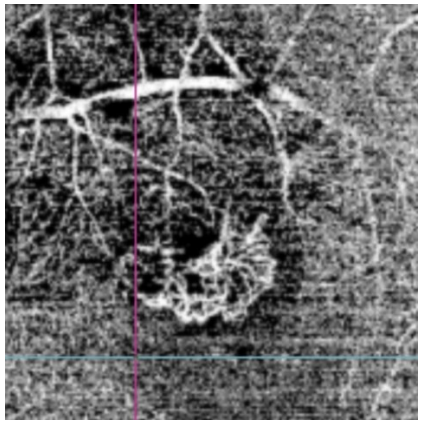
Cas du Dr Sarah Mrejen XV-XX

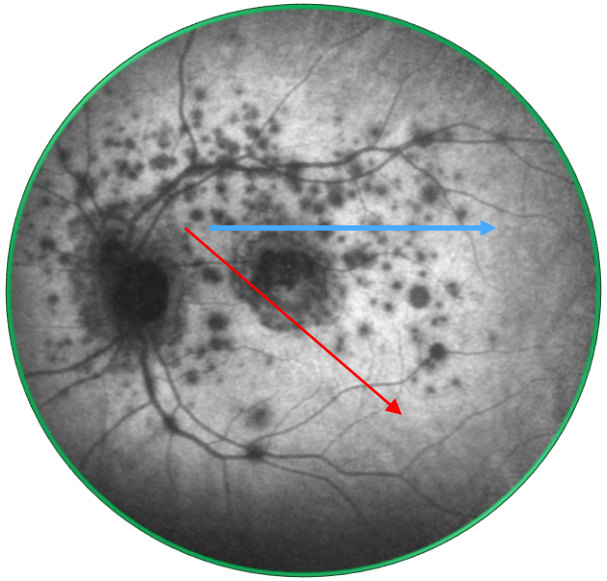
MEWDS secondaire à des NVC

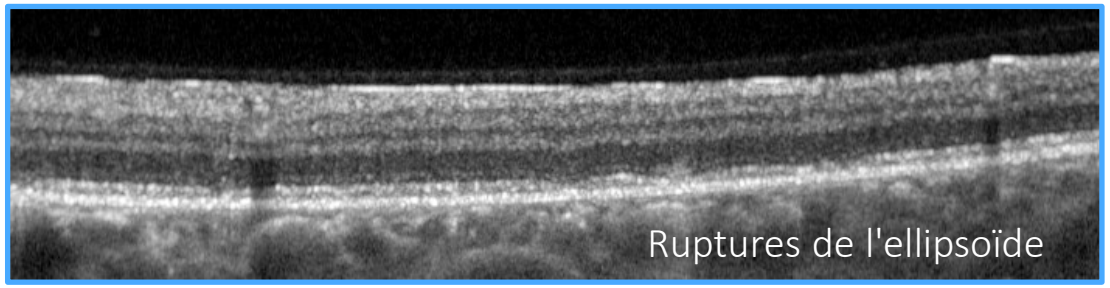


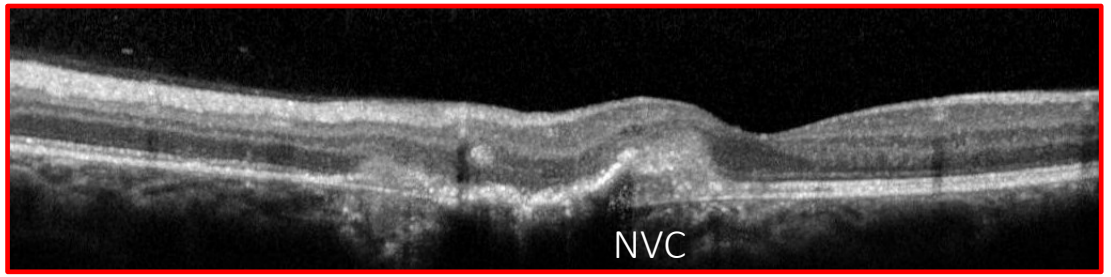












- 2 ans plus tard , nouvelle récidive de NVC
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Cas du Dr Sarah Mrejen XV-XX

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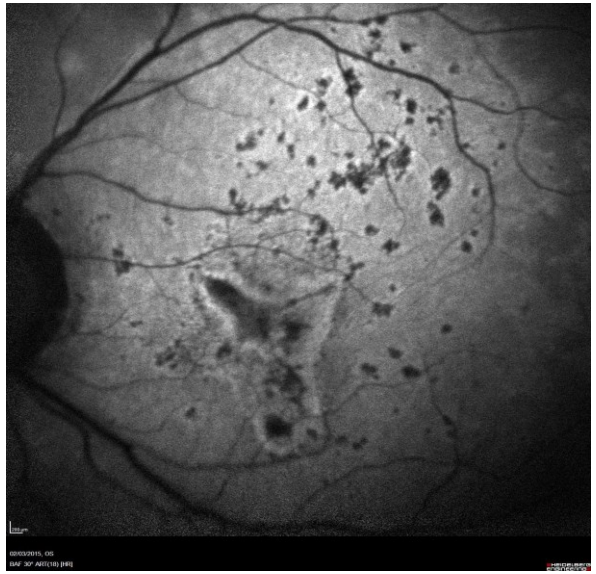

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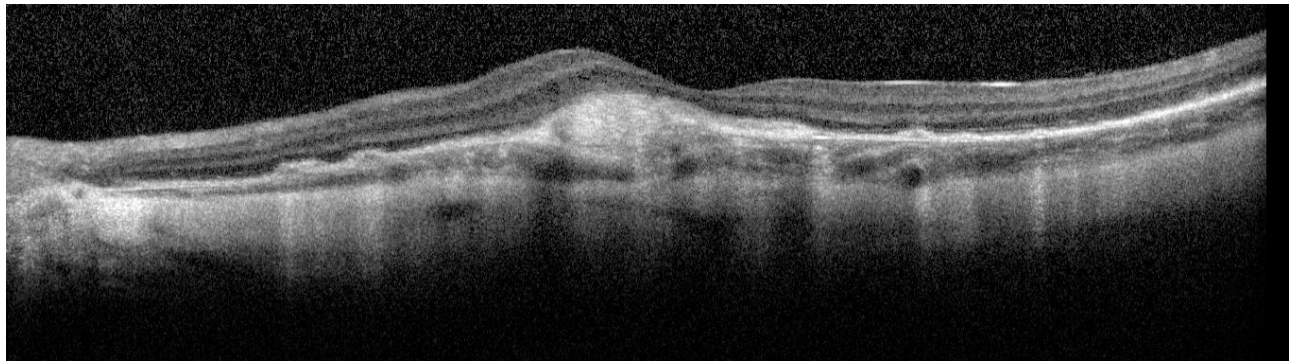

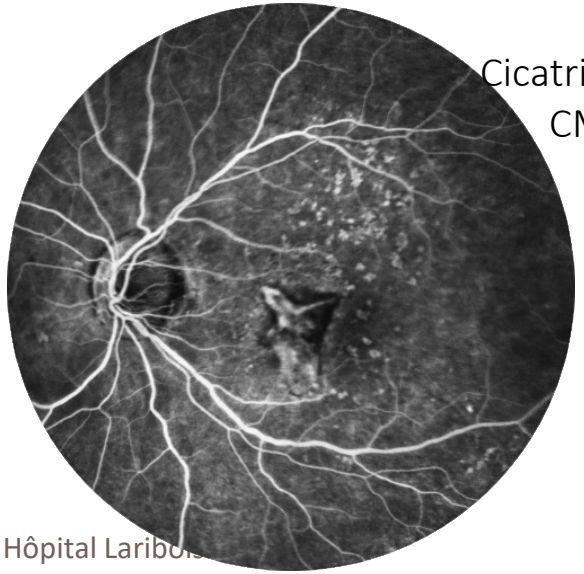

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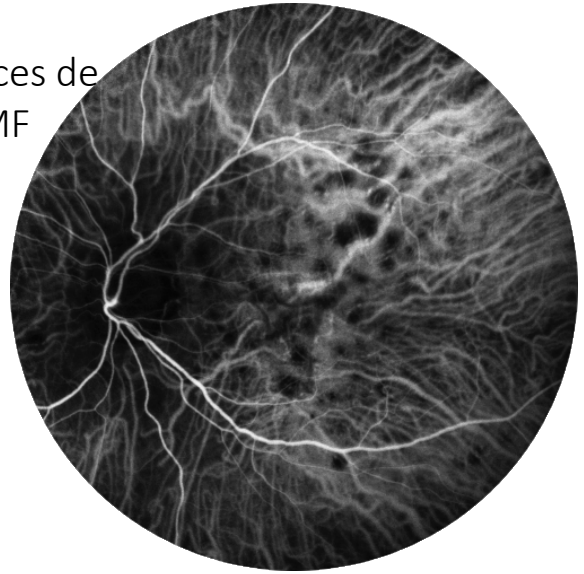
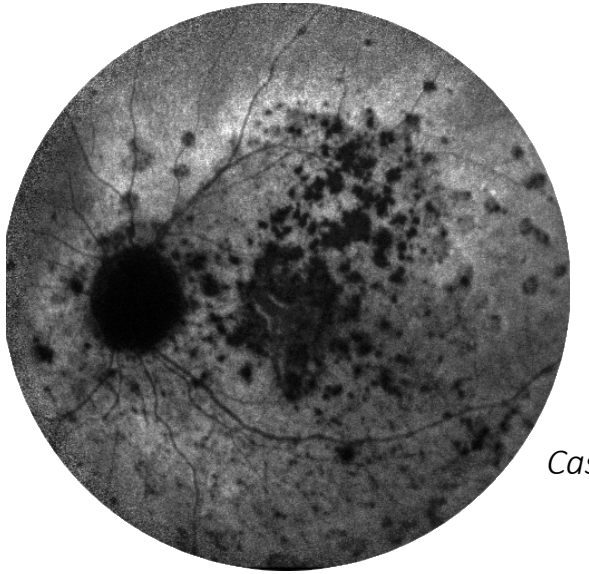
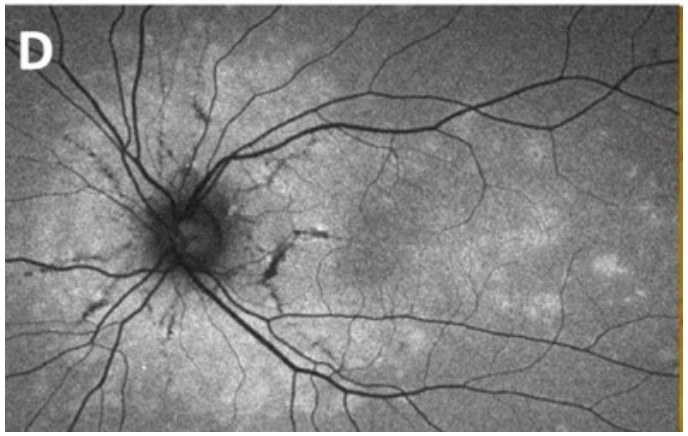


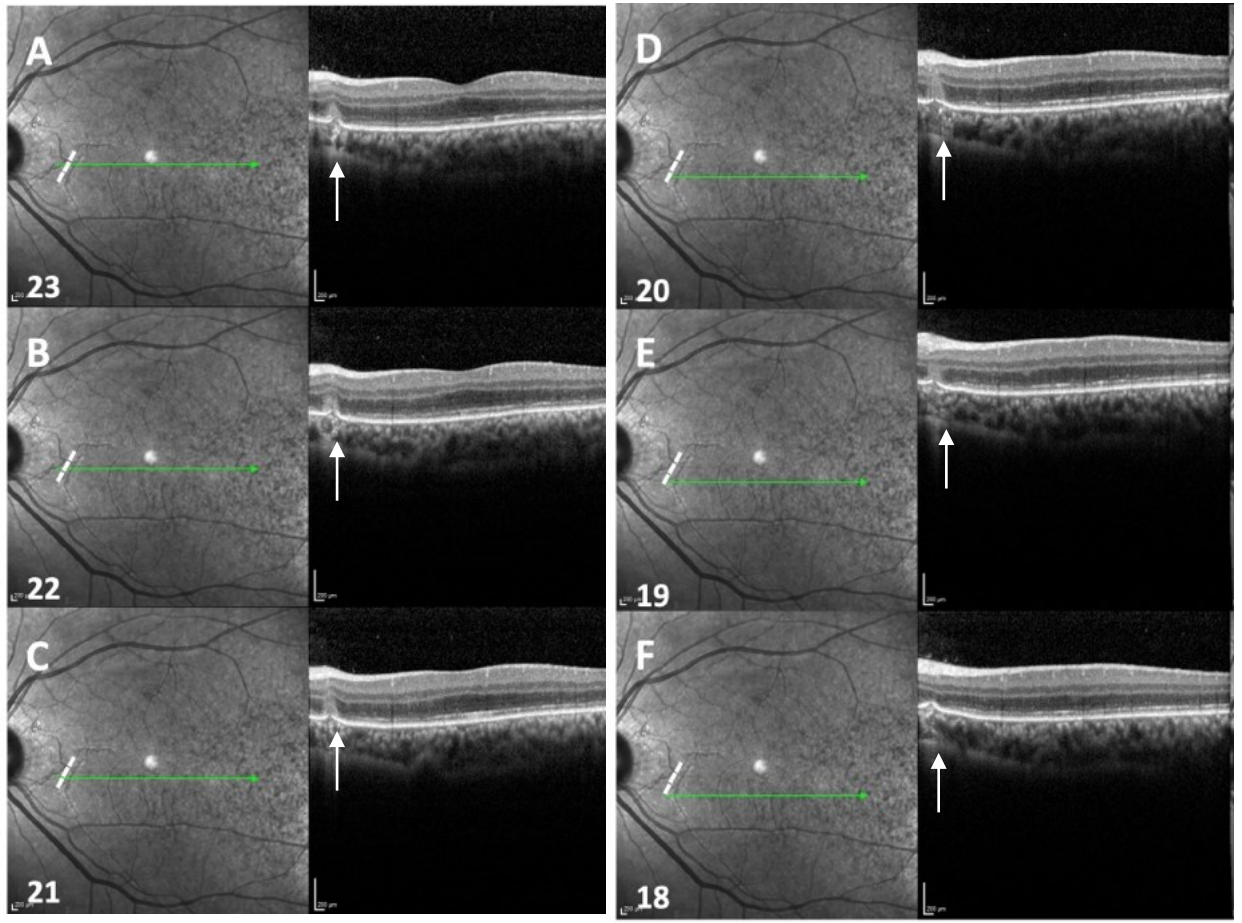


1 an plus tard
ICGA 30 mn

Cas du Dr MH Errera XV-XX, Pittsburg

Stries angioïdes et MEWDS





JAMA Ophthalmology | Original Investigation

Acute Retinopathy in Pseudoxanthoma Elasticum

Martin Gliem, MD; Johannes Birtel, MD; Philipp L. Müller, MD; Doris Hendig, PhD; Isabel Faust, PhD; Philipp Herrmann, MD, PhD; Frank G. Holz, MD; Grazyna Adamus, PhD; Peter Charbel Issa, MD, DPhil

Jama Ophthalmol. 2019;137(10):1165-1173.

RECURRENCE OF ACUTE RETINOPATHY IN PSEUDOXANTHOMA ELASTICUM

Ramtohul, Prithvi MD[1]; Cabral, Diogo MD[1]; Cicinelli, Maria Vittoria MD[2,3]; Freund, K. Bailey MD[1,4]

Retinal Cases & Brief Reports ():10.1097/ICB.0000000000001363, November 03, 2022.

Université Paris Cité

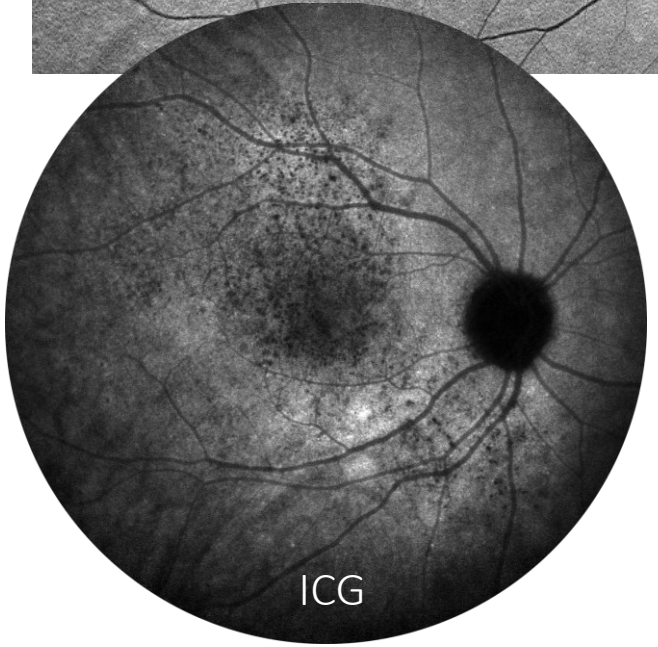
MEWDS, Diagnostic différentiel

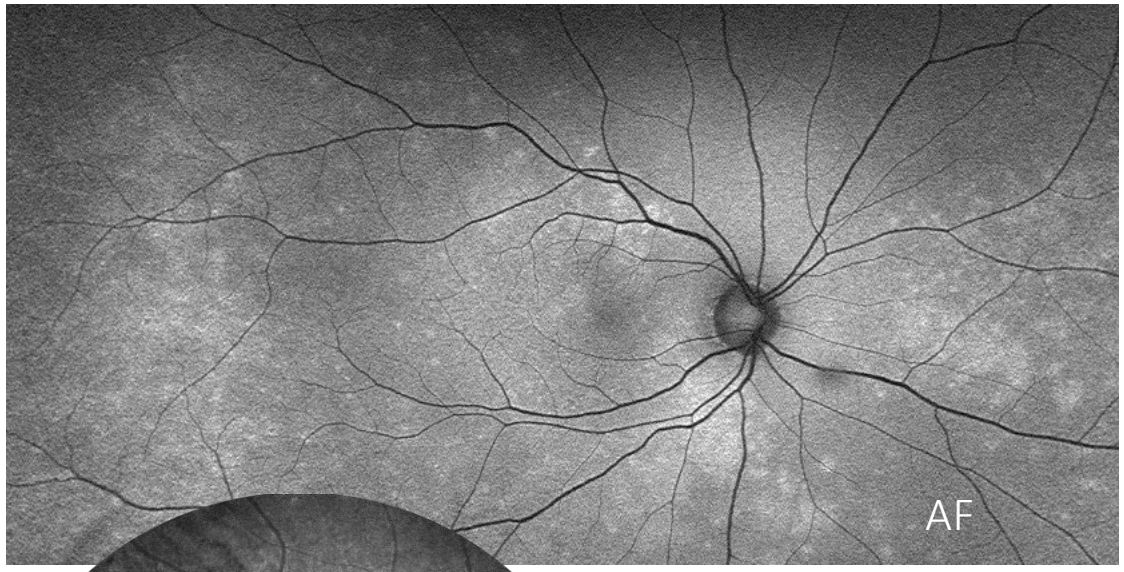
Masqueraders of multiple evanescent white dot syndrome (MEWDS)

Jonathan F. Russell · Francesco Pichi · Nathan L. Scott · Matthew J. Hartley · Dugald Bell · Aniruddha Agarwal · Belinda Leong · Gary N. Holland · K. Bailey Freund · David Sarraf

Int Ophthalmol. 2020;40(3):627-638.

- Syphilis
- Lymphome oculaire
- Choroidite multifocale
- Sarcoidose
- Tuberculose
- CAR
- AZOOR





Syphilis

À quel niveau cellulaire se trouve l'atteinte du MEWDS?

Multiple Evanescent White Dot Syndrome

Nicole E. Gross, MD; Lawrence A. Yannuzzi, MD; K. Bailey Freund, MD; Richard F. Spaide, MD; Gian Paolo Amato, MD; Ruth Sigal, MD

Arch Ophthalmol. 2006;124:493-500

cent lesions. Based on the angiographic findings, it seems as if MEWDS is a chorioretinopathy with varying degrees of retinal and choroidal involvement.

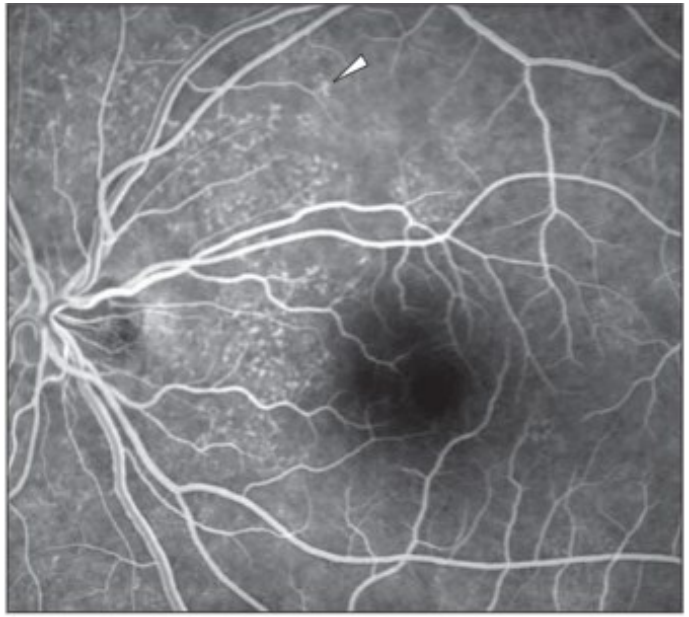


Figure 2. Hyperfluorescent dots on an early fluorescein angiogram demonstrating window defects and in the retinal vasculature (arrowhead).

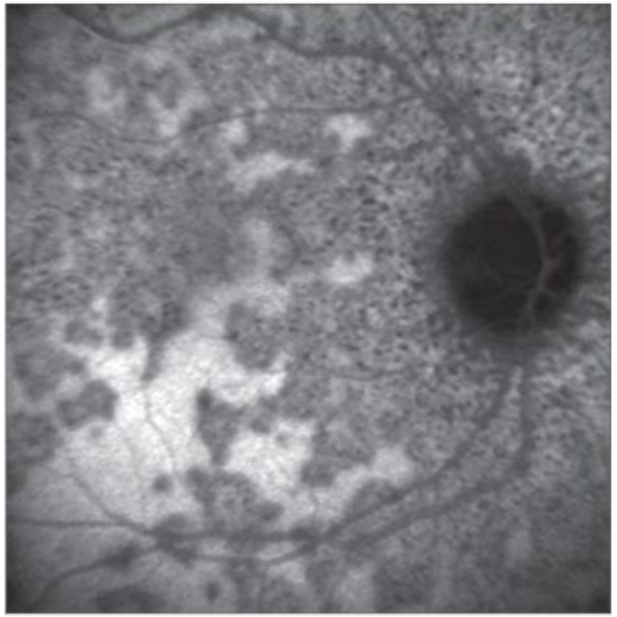


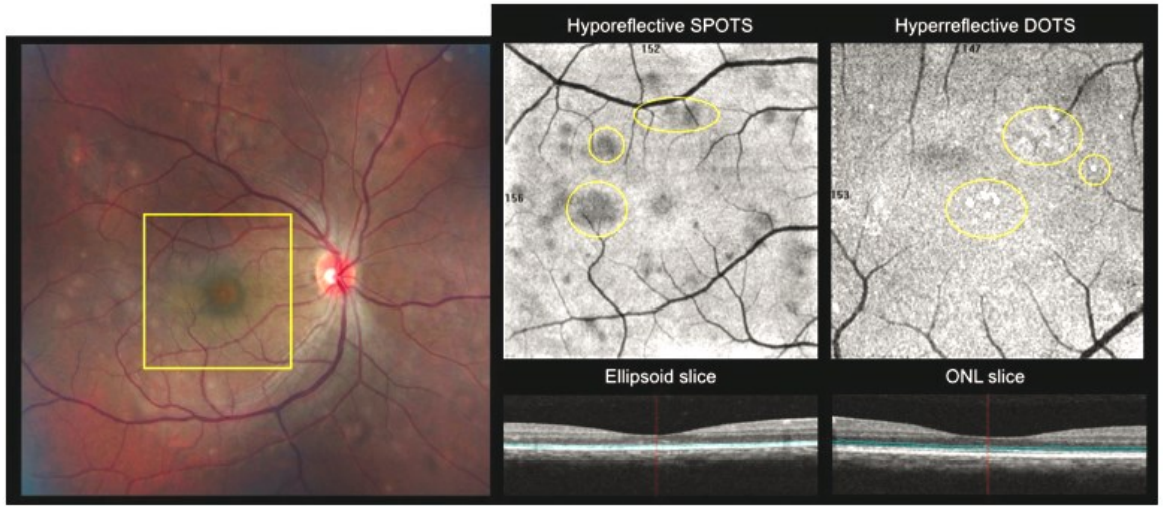
Figure 4. Indocyanine green angiogram demonstrating hypofluorescent dots and spots with a predilection for the area around the optic nerve.

EN FACE OPTICAL COHERENCE TOMOGRAPHY AND OPTICAL COHERENCE TOMOGRAPHY ANGIOGRAPHY OF MULTIPLE EVANESCENT WHITE DOT SYNDROME

New Insights Into Pathogenesis

Pichi F, Srvivastava SK, Bailey Feund K, Sarraf D,, et al. Retina 2016;36:S178-S188.

In conclusion, MEWDS seems to be primarily a disease of the RPE and photoreceptor inner and outer segment complex. Its evanescent nature suggests that



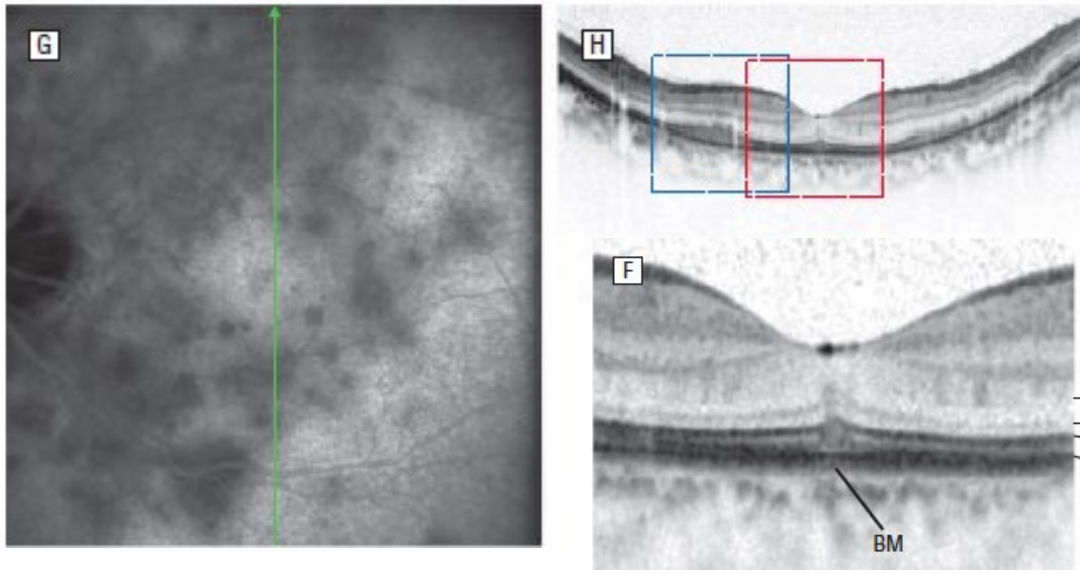
MEWDS , Epithélium Pigmentaire et Photorécepteurs

CLINICAL SCIENCES

Features and Function of Multiple Evanescent White Dot Syndrome

Masanori Hangai, MD; Masahiro Fujimoto, MD; Nagahisa Yoshimura, MD

Arch Ophthalmol. 2009;127(10):1307-1313



Conclusion: Enhanced SD-OCT revealed abnormalities in the photoreceptor layer that were specific to MEWDS and that, with retinal shape and function, seemed to change location during clinical recovery from MEWDS.

L'OCT a révélé des anomalies dans la couche des photorécepteurs qui étaient spécifiques du MEWDS

are thought to involve RPE abnormalities. Therefore, our results support the assumption that the MEWDS disease process occurs in the RPE. However, our results do not provide direct evidence of involvement of the choroid in the disease process or determine whether the choroid and RPE are ischemic in MEWDS.

nos résultats soutiennent l'hypothèse selon laquelle le processus du MEWDS se produit dans l'EP

EXPANDED CLINICAL SPECTRUM OF MULTIPLE EVANESCENT WHITE DOT SYNDROME WITH MULTIMODAL IMAGING

MARCELA MARSIGLIA, MD, PHD,*† ROBERTO GALLEGO-PINAZO, MD, PHD,‡
EDUARDO CUNHA DE SOUZA, MD,§ MARION R. MUNK, MD,¶** SUQUIN YU, MD,††
SARAH MREJEN, MD,‡‡ EMMETT T. CUNNINGHAM, JR., MD, PHD,§§ BRANDON LUJAN, MD,¶¶
NAOMI R. GOLDBERG, MD, PHD,*** THOMAS A. ALBINI, MD,††† ALAIN GAUDRIC, MD,‡‡‡
CATHERINE FRANCAIS, MD,§§§ RICHARD B. ROSEN, MD,¶¶¶ K. BAILEY FREUND, MD,*†****
LEE M. JAMPOL, MD,¶ LAWRENCE A. YANNUZZI, MD*†*****

RETINA. 2016;36(1):64-74.

" There was no evidence of primary RPE disease "

"Multiple evanescent white dot syndrome seems as a primary reversible nondestructive retinal pigment epitheliopathy "

WHY THE DOTS ARE BLACK ONLY IN THE LATE PHASE OF THE INDOCYANINE GREEN ANGIOGRAPHY IN MULTIPLE EVANESCENT WHITE DOT SYNDROME

Alain Gaudric, MD,* Sarah Mrejen, MD†

Retinal Cases and Brief Reports 2017

Purpose: To determine what retinal layer is primarily involved in multiple evanescent white dot syndrome compared with acute posterior multifocal placoid pigment epitheliopathy using multimodal imaging including indocyanine green angiography.

Methods: Color fundus photographs and fluorescein angiography, ICGA and spectral domain optical coherence tomography images of two typical acute cases, one of acute posterior multifocal placoid pigment epitheliopathy and one of multiple evanescent white dot syndrome, were assessed and compared.

Results: In both diseases, white plaques or dots were seen at the posterior pole. The hypofluorescence of acute posterior multifocal placoid pigment epitheliopathy in the early phase of the fluorescein angiography and throughout the ICGA sequence suggested a multifocal choroidal hypoperfusion as the cause of the opacification of the outer retina corresponding to its hyper-reflectivity on spectral domain optical coherence tomography. In multiple evanescent white dot syndrome, there was no choroidal filling defect and the white spots were silent in the early phase of the ICGA, whereas they became black only in the late phase. The outer retina was also altered at the spots.

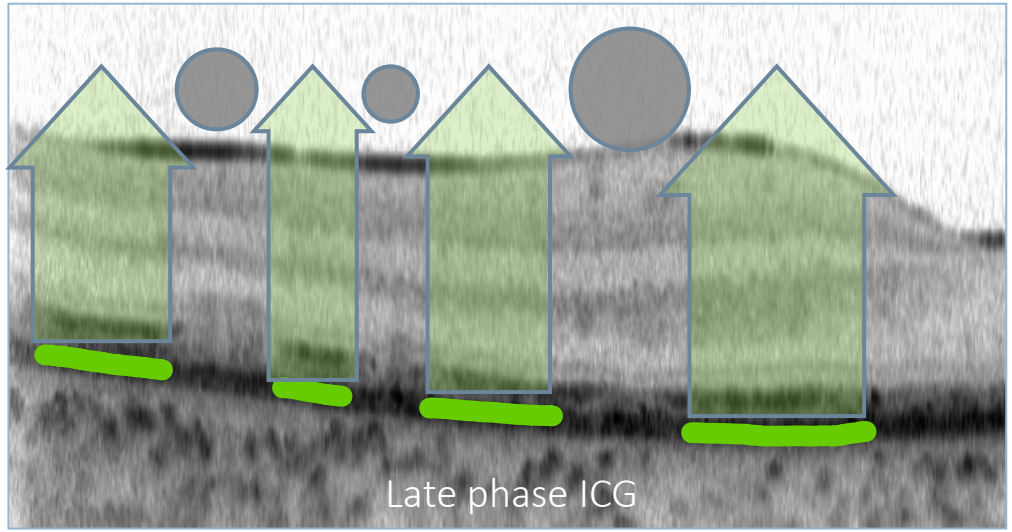
Conclusion: In multiple evanescent white dot syndrome, the late hypofluorescence of the white spots on ICGA was likely due to the absence of ICG uptake by a damaged retinal pigment epithelium. Multiple evanescent white dot syndrome seems as a primary reversible nondestructive retinal pigment epitheliopathy, whereas acute posterior multifocal placoid pigment epitheliopathy is a true choroidopathy.

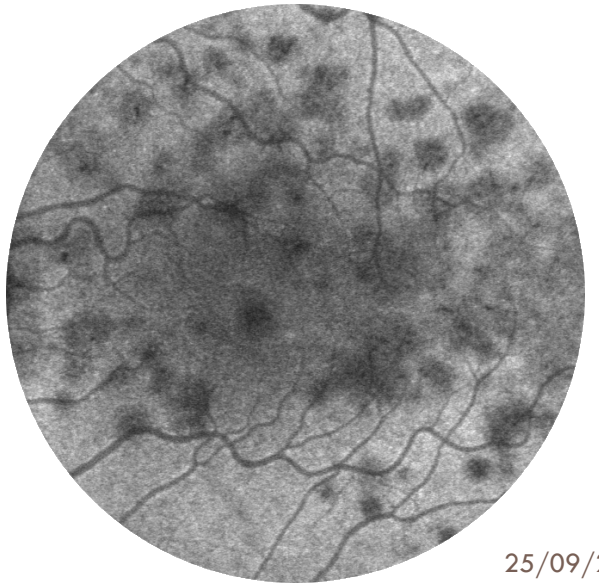
RETINAL CASES & BRIEF REPORTS 11:S81–S85, 2017

Interprétation de la phase tardive de l'ICG

- Dans le MEWDS
  - L'EP ne parvient pas à internaliser l'ICG présent dans la choroide au niveau des taches blanches
  - Par contraste avec l'EP avoisinant les taches, deviennent progresivement sombre
  - Ce dysfonctionnement de l'EP n'implique pas de mort cellulaire mais pertube transitoirement le fonctionnement des photorécepteurs

MEWDS





- *Owens SL. Brit J Ophthalmol. 1996;80(3):263*
- *Gaudric A, Mrejen S. Retin Cases Brief Reports. 2016;11*
- *Zicarelli F, Ocular immunology and inflammation. 2019:1-7*

Critères de diagnostic : appréciation critique

Classification Criteria For Multiple Evanescent White Dot Syndrome

THE STANDARDIZATION OF UVEITIS NOMENCLATURE (SUN) WORKING GROUP[1,2,3,4,*]

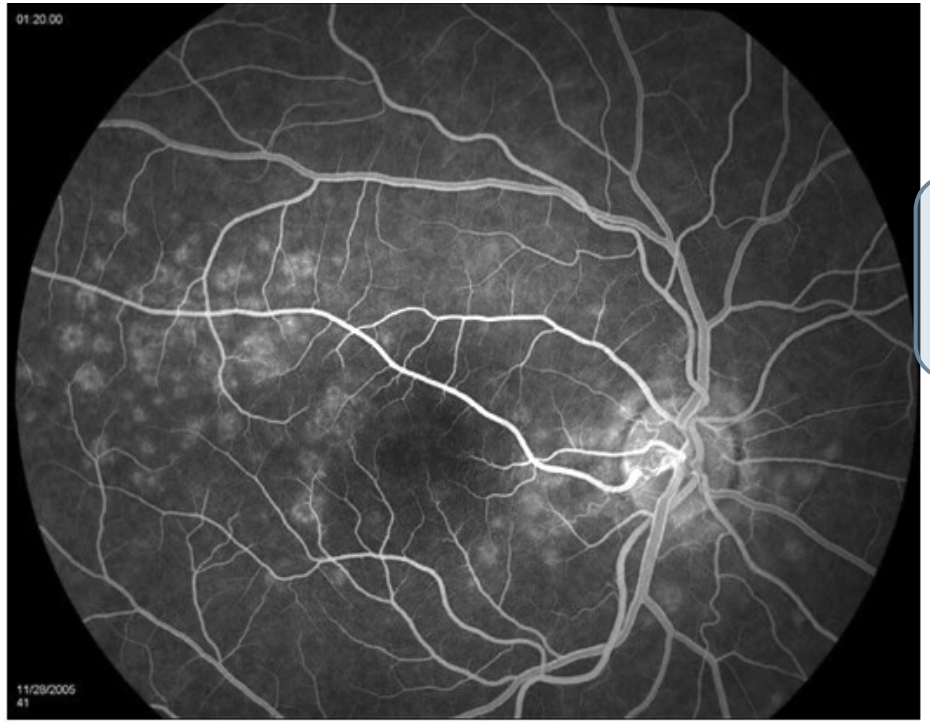


FIGURE 2. Fluorescein angiogram of a case of multiple evanescent white dot syndrome, demonstrating the "wreath-like" nature of the chorioretinal lesions.

Aucune mention de l'ICG

TABLE 2. Classification Criteria for Multiple Evanescent White Dot Syndrome

Criteria

1. Multifocal chorioretinal gray-white spots with foveal granularity

AND

2. Characteristic fluorescein angiogram or optical coherence tomogram (OCT)
   a. "Wreath-like" hyperfluorescent lesions on fluorescein angiogram OR
   b. Hyperreflective lesions on OCT extending from the retinal pigment epithelium, into and/or through the ellipsoid zone into the outer nuclear layer of the retina

AND

3. Absent to mild anterior chamber and vitreous inflammation

Exclusions

1. Positive serologic test for syphilis using a treponemal test
2. Evidence of sarcoidosis (either bilateral hilar adenopathy on chest imaging or tissue biopsy demonstrating non-caseating granulomata)
3. Bilateral simultaneous disease onset

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Merci de votre attention

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Service d'Ophtalmologie
Hôpital Lariboisière

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RETINAL CASES & BRIEF REPORTS 11:S81–S85, 2017

Multimodal Imaging of Multiple Evanescent White Dot Syndrome: A New Interpretation

Federico Zicarelli, Alessandro Mantovani, Chiara Preziosa & Giovanni Staurenghi

Ocul Immunol Inflamm. August 2019:1-7.

To conclude, MEWDS etiology and pathogenesis are still debated. Multimodal imaging allowed us to suggest the RPE to be the key tissue involved in the inflammatory process. The photoreceptors alterations may be consequent to the RPE dysfunction rather than the expression of a direct damage induced to the cells by the disease. Further studies are needed to confirm our hypothesis.

L'imagerie multimodale nous a permis de suggérer que l'EPR est le tissu clé impliqué dans le processus inflammatoire. Les altérations des photorécepteurs peuvent être la conséquence du dysfonctionnement de l'EPR plutôt que l'expression d'un dommage direct induit aux cellules par la maladie.

APMPPE

MEWDS

Aucun conflit d'intérêt

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Merci de votre attention

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