



FACULTÉ DE
MÉDECINE
& MAÏEUTIQUE



ICGA is still relevant today

Cas clinique n°5

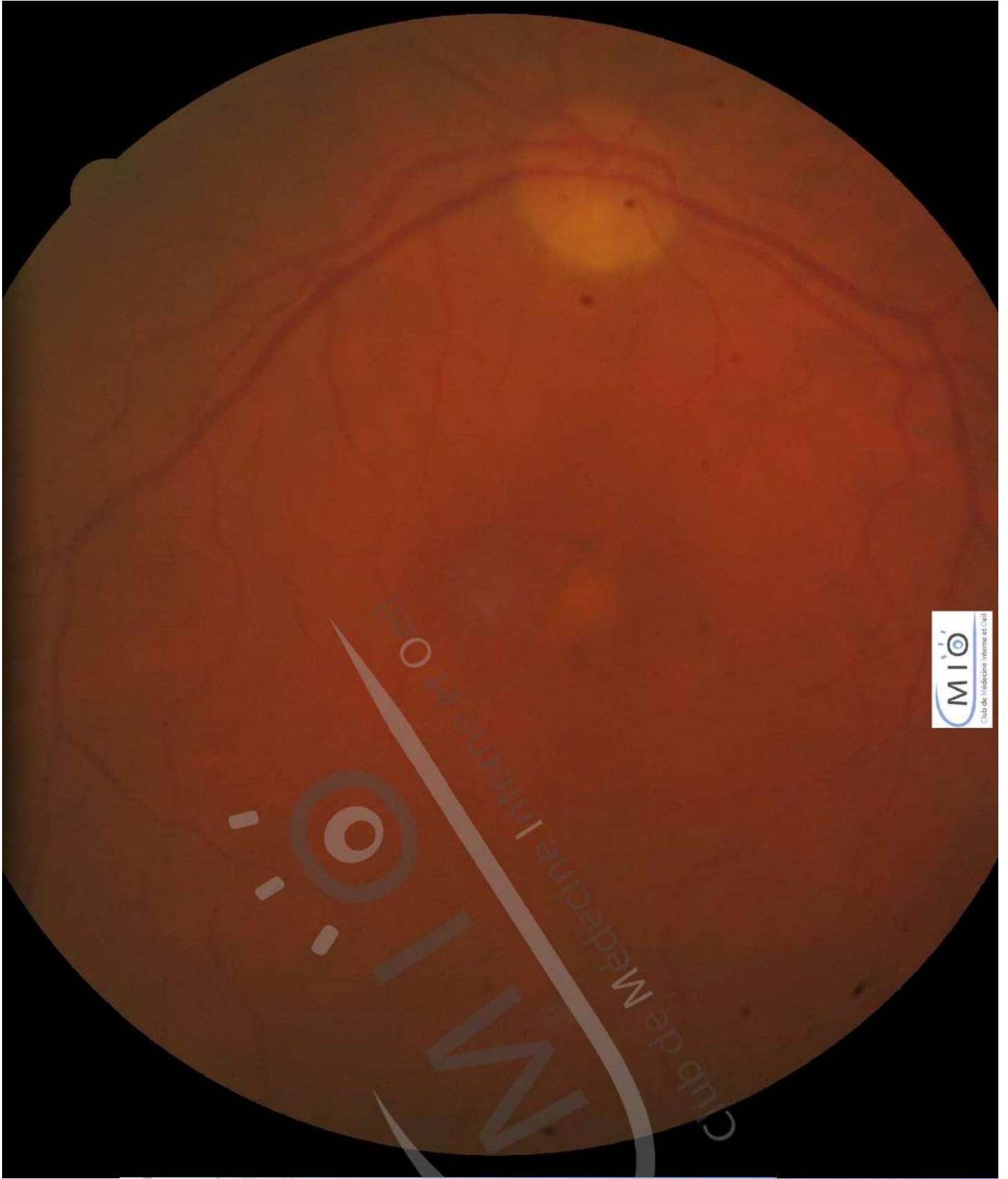
FMC du 13 octobre 2017

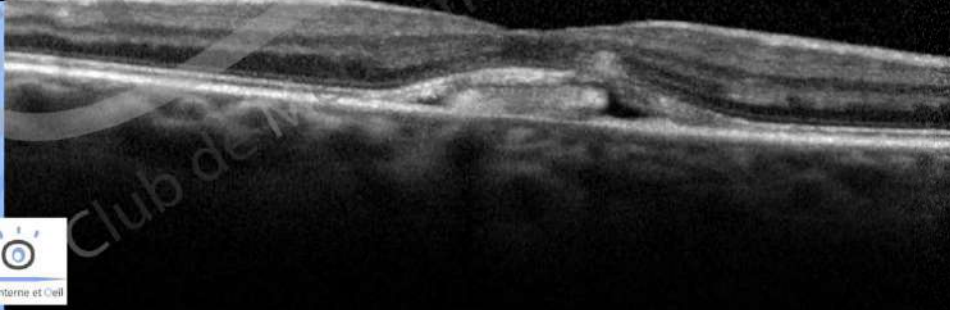
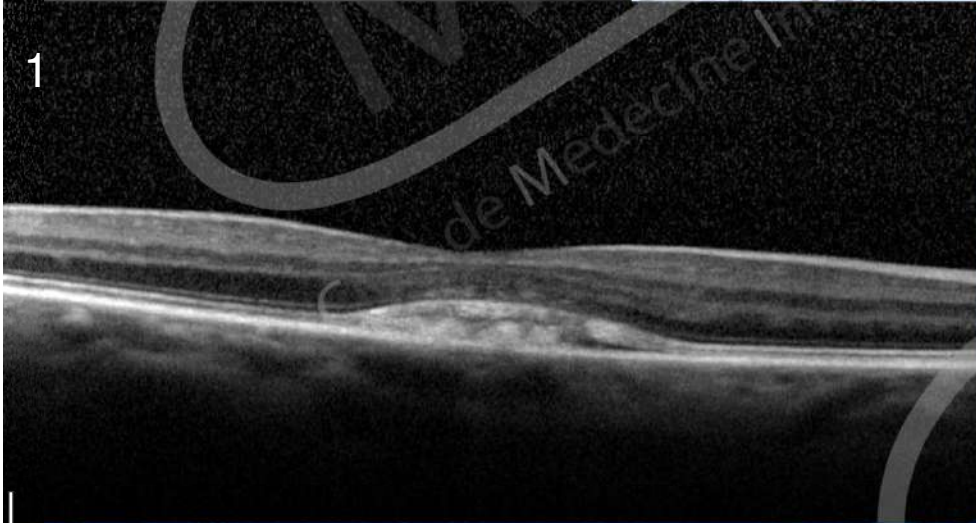
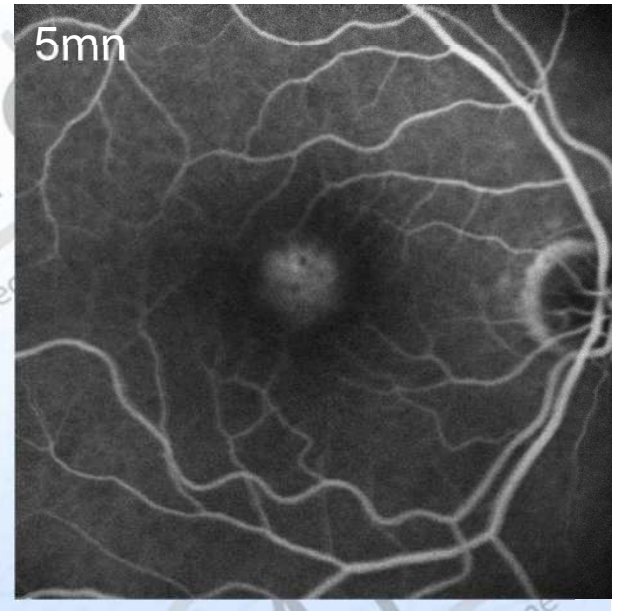
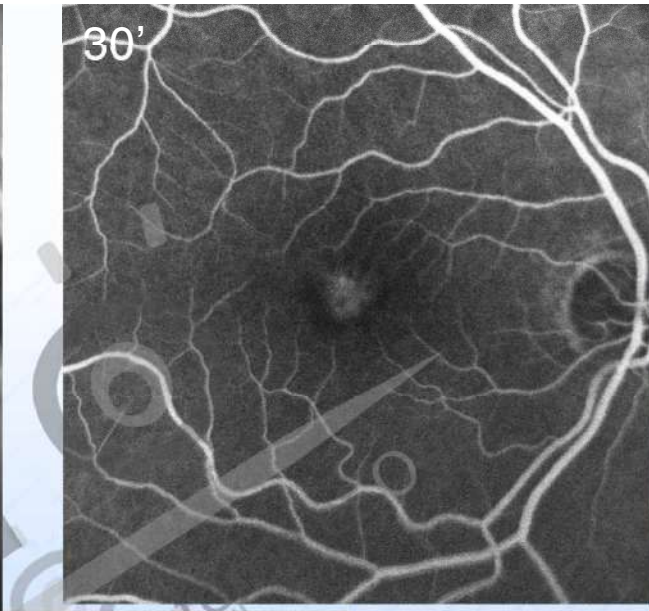
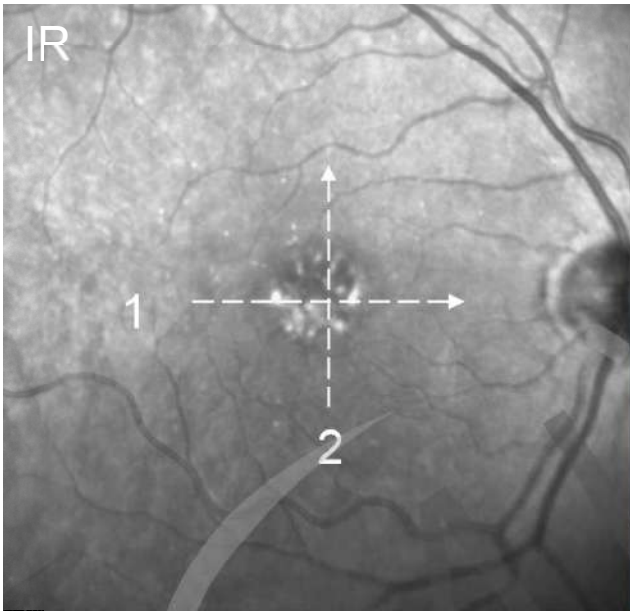
TRAN THC

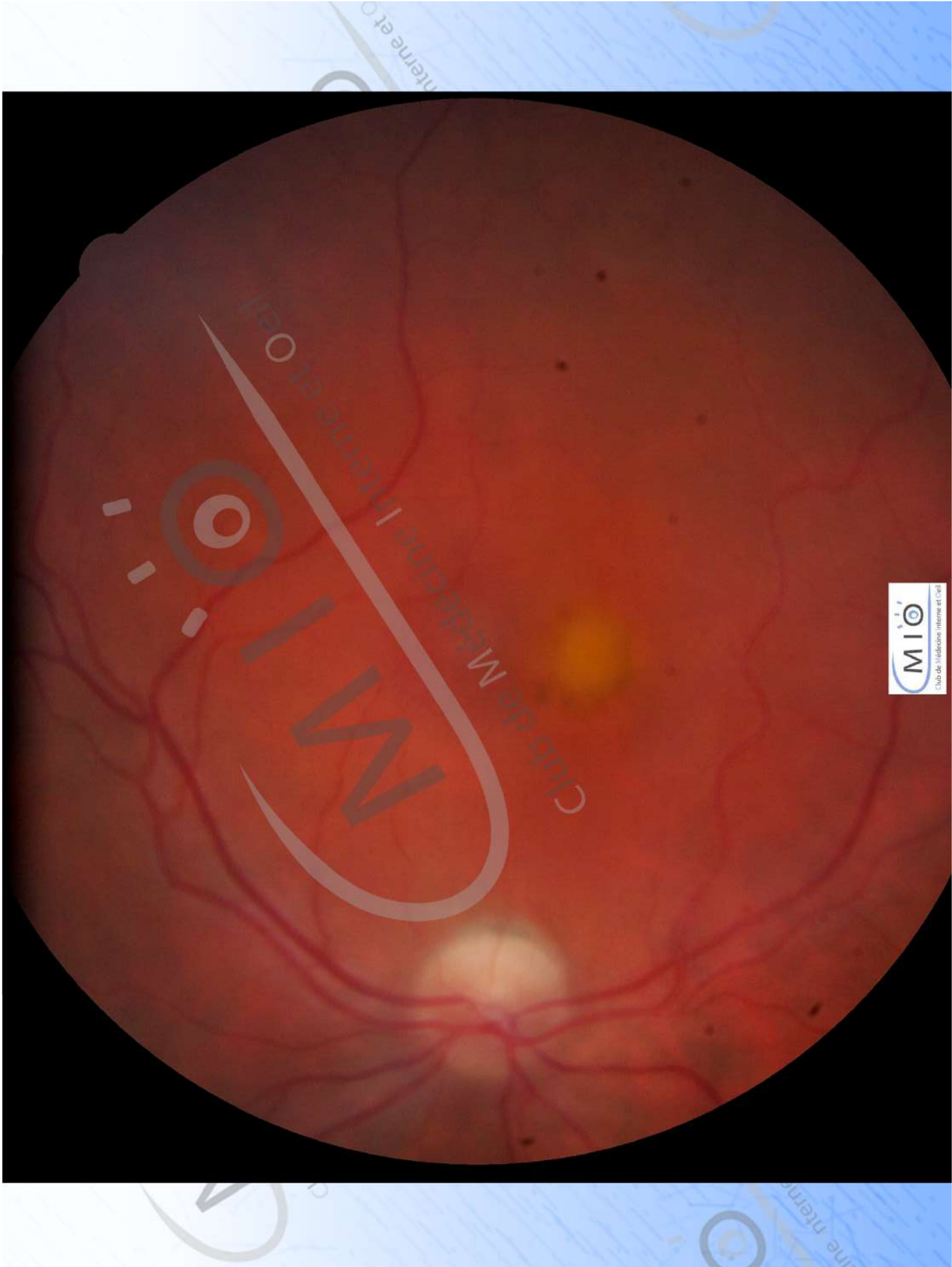
Email : tran.hachau@ghicl.net

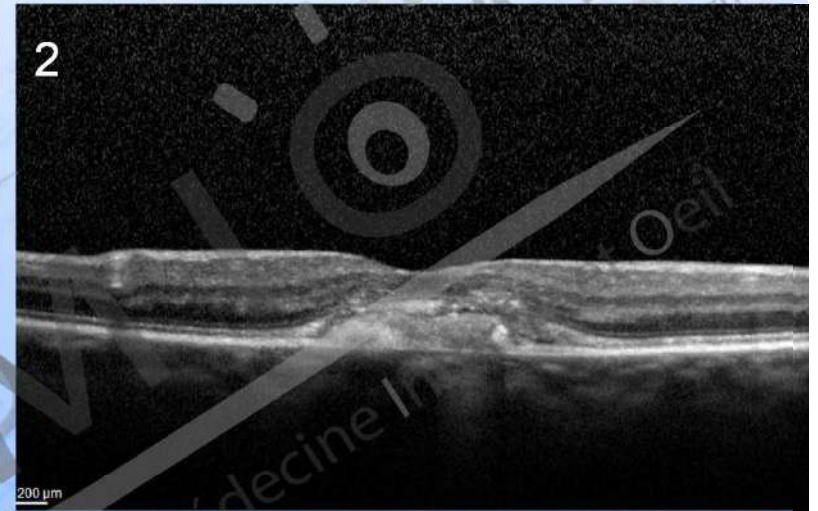
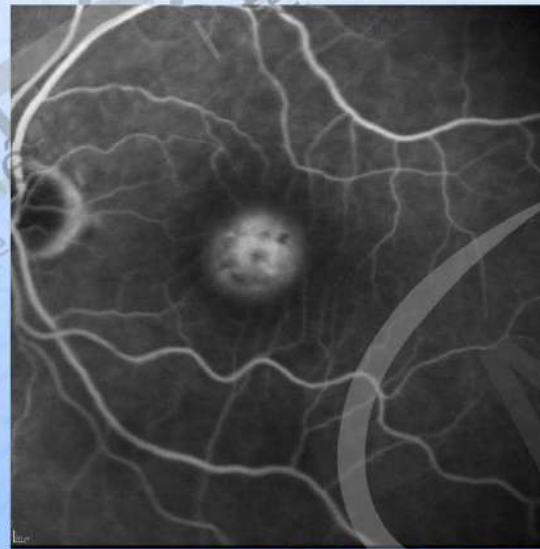
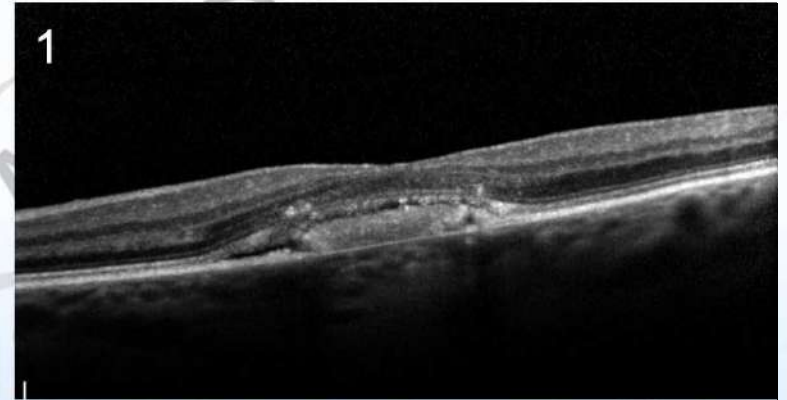
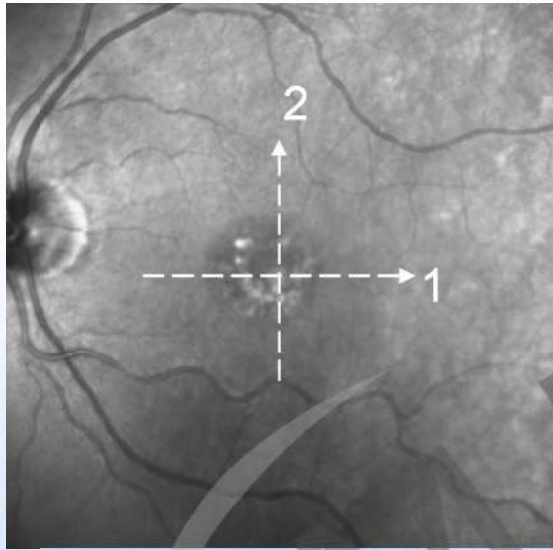
***Groupe des hôpitaux Catholique de Lille
France***

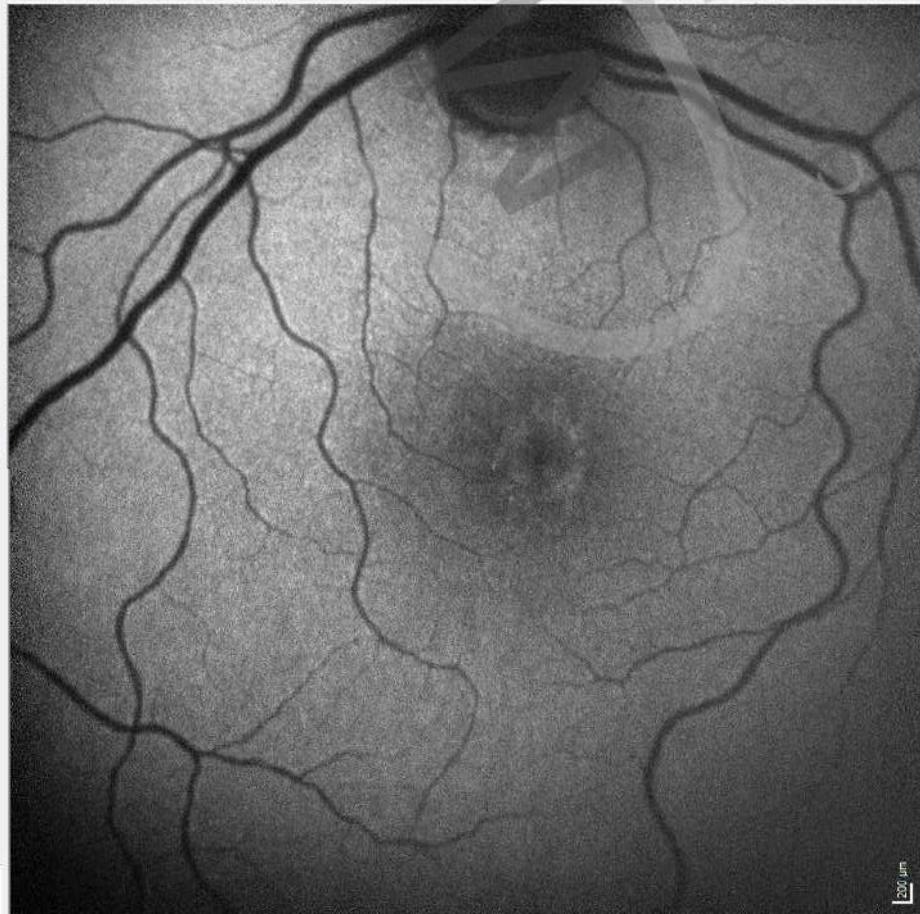
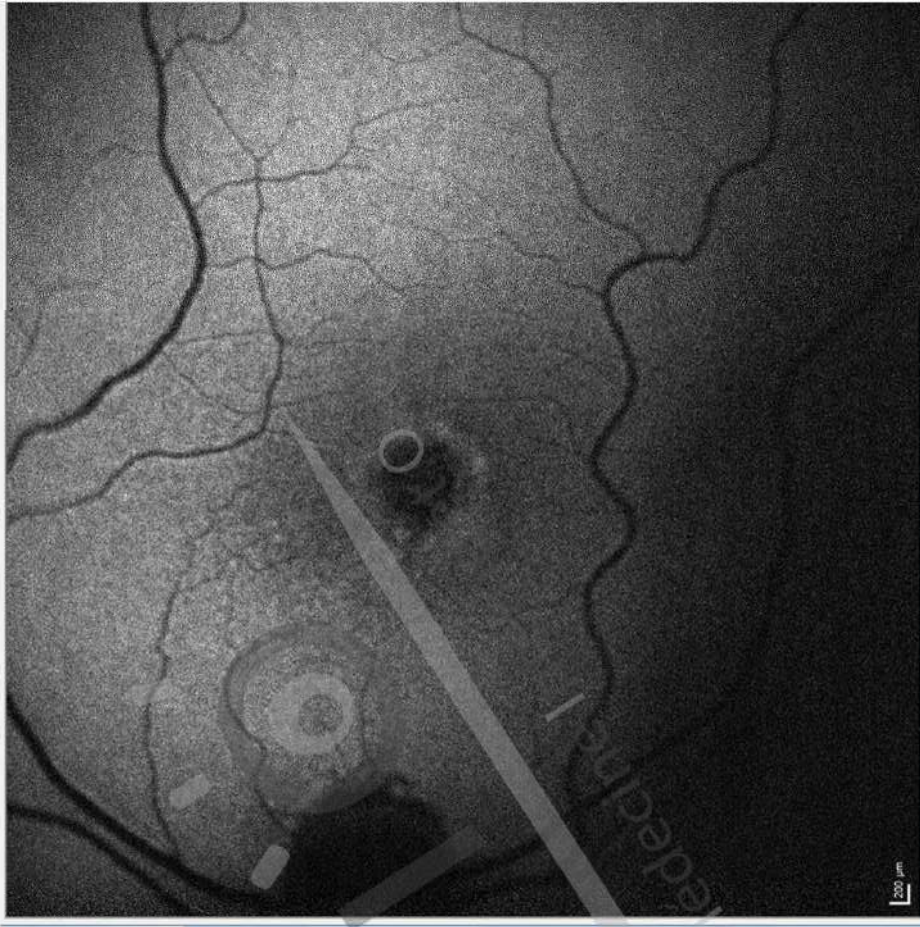
- 2010
- 75 year-old woman
- VA
 - OD 20/50
 - OS 20/60
- Slit- lamp examination unremarquable
 - OD : Cataract
 - OS : IOL
- Funduscopy
 - Vitreous : clear
 - Peripheral retina : unremarkable











Diagnosis ?

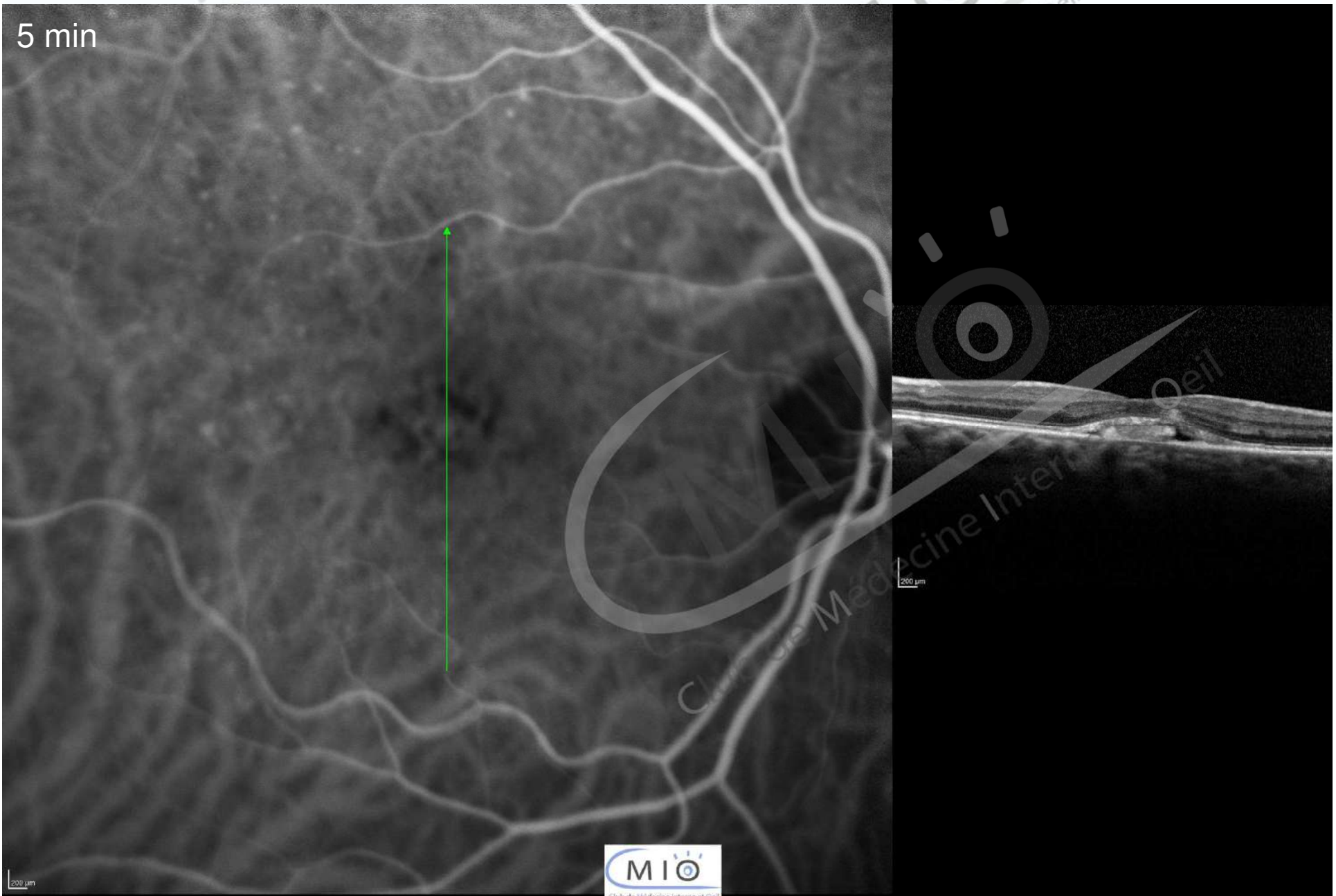
1. Adult Vitelliform Macular Degeneration

2. Neovascular AMD

1 min

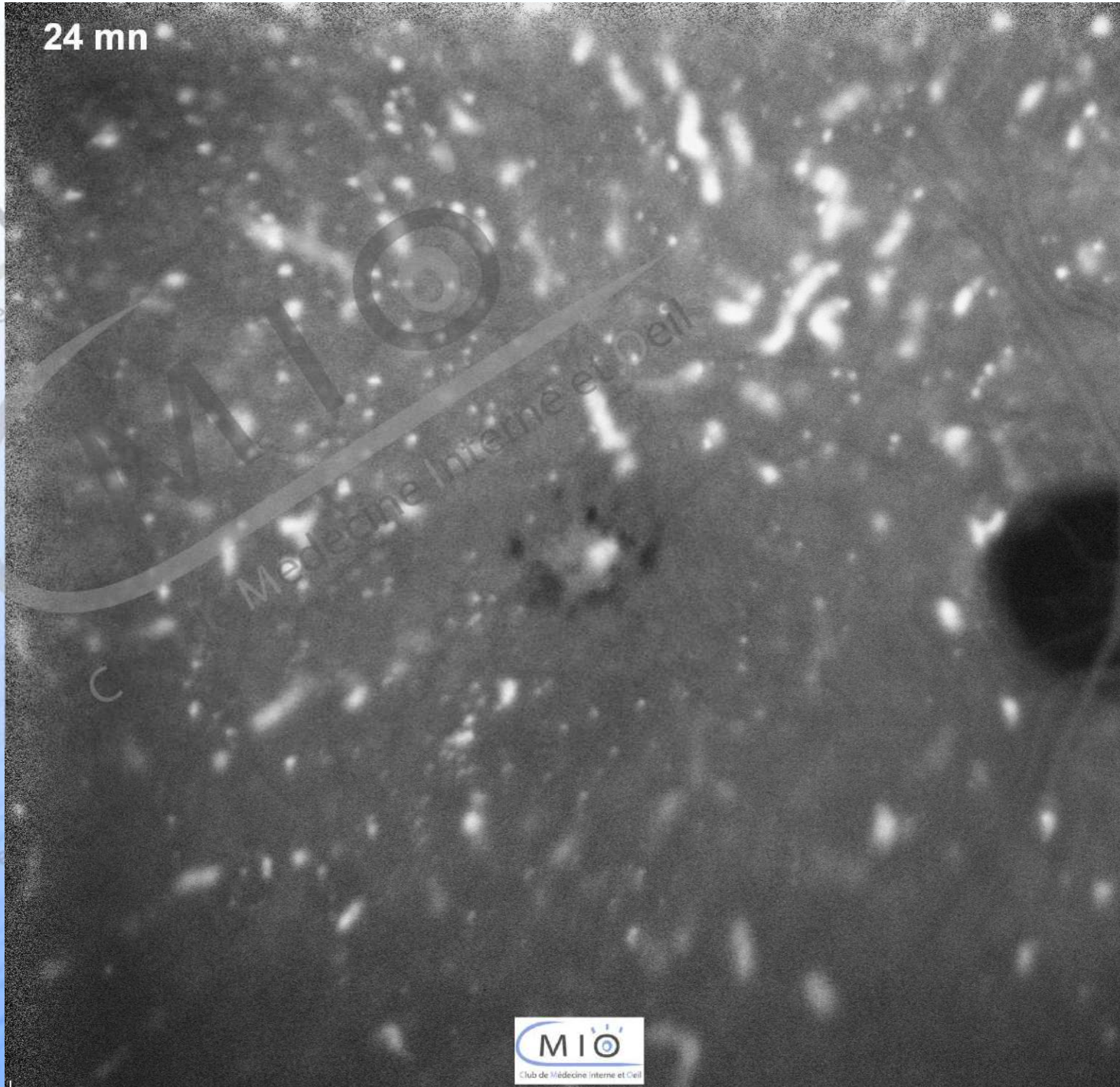


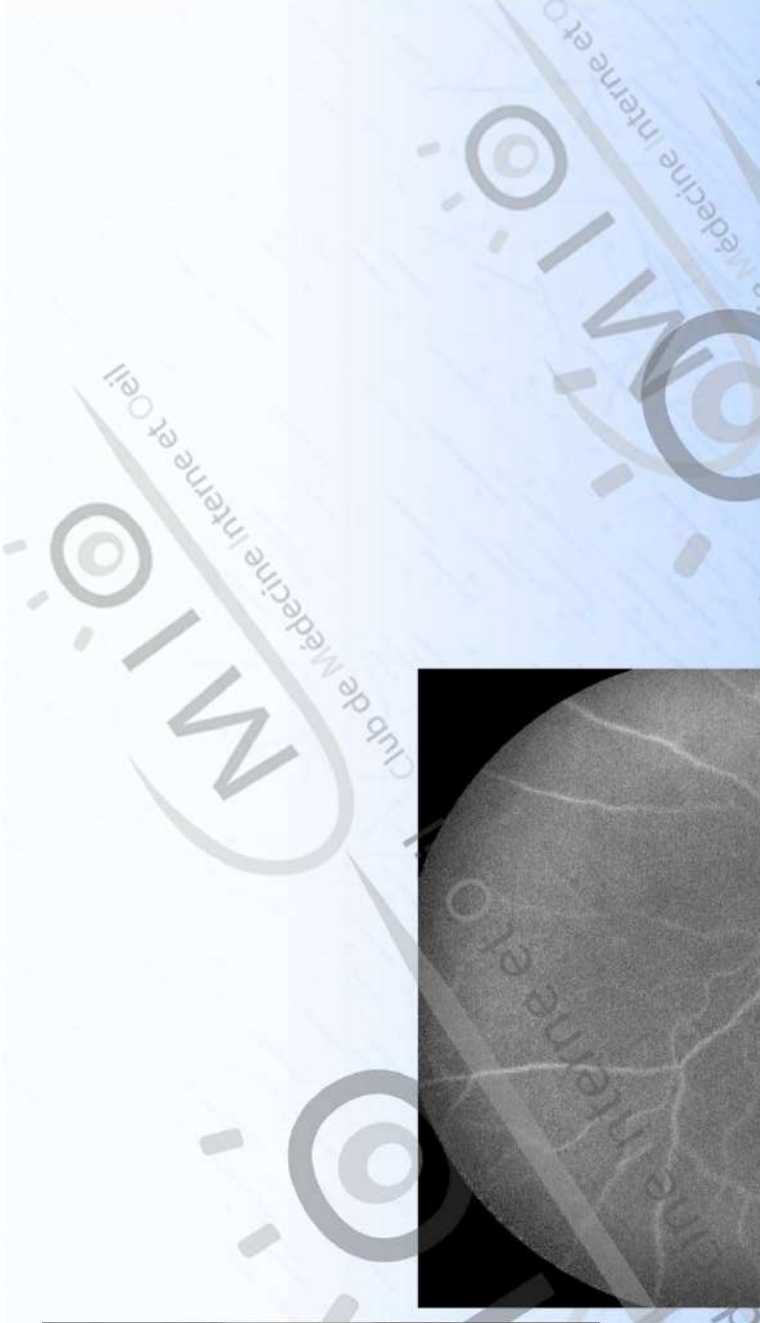
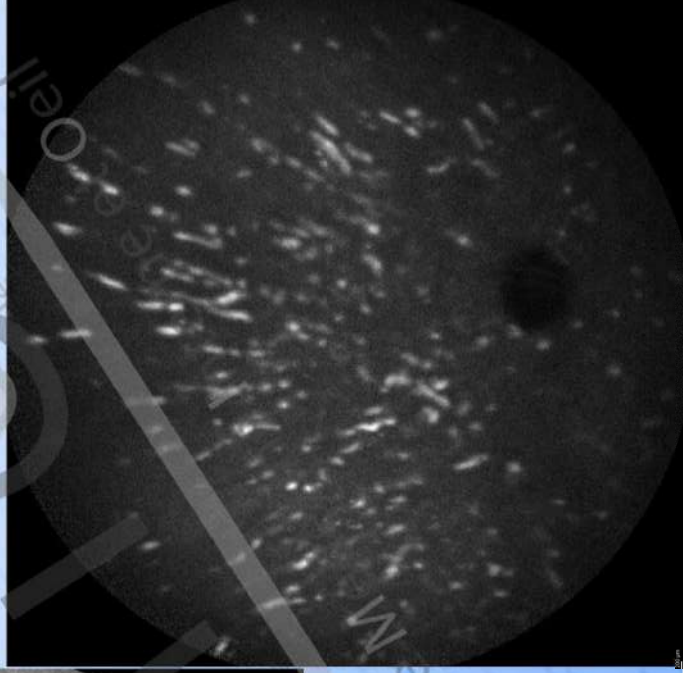
5 min

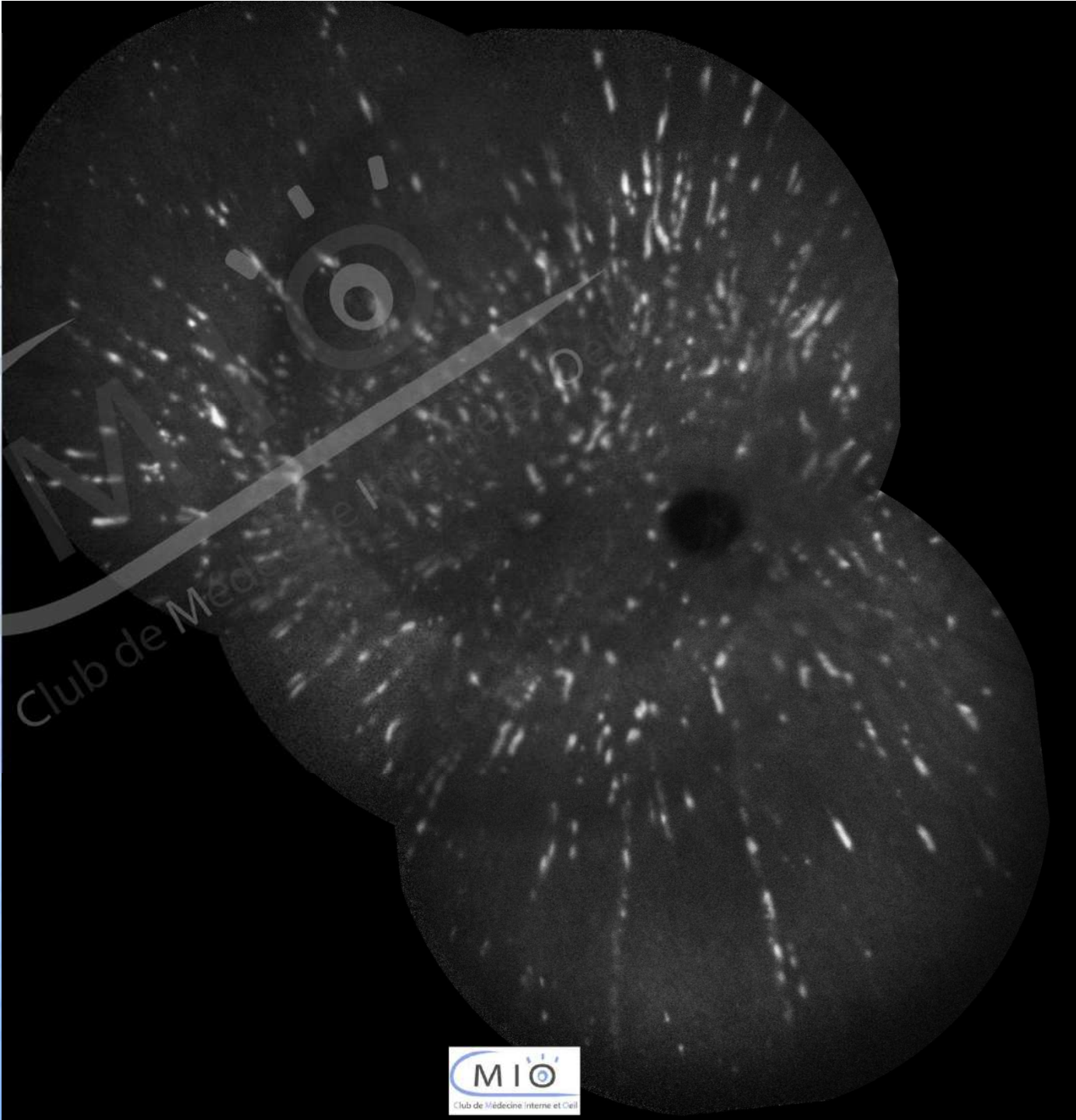


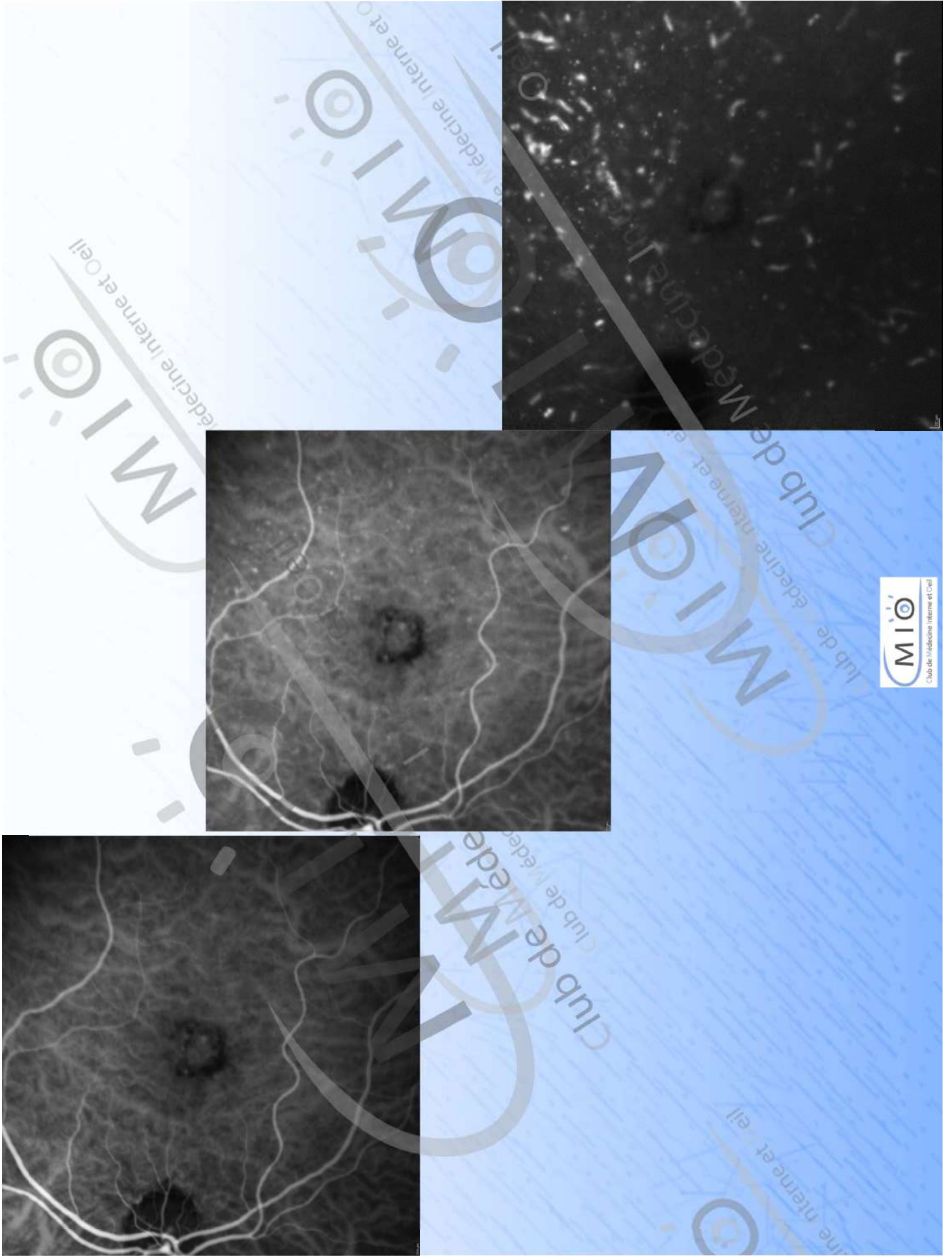
200 μm

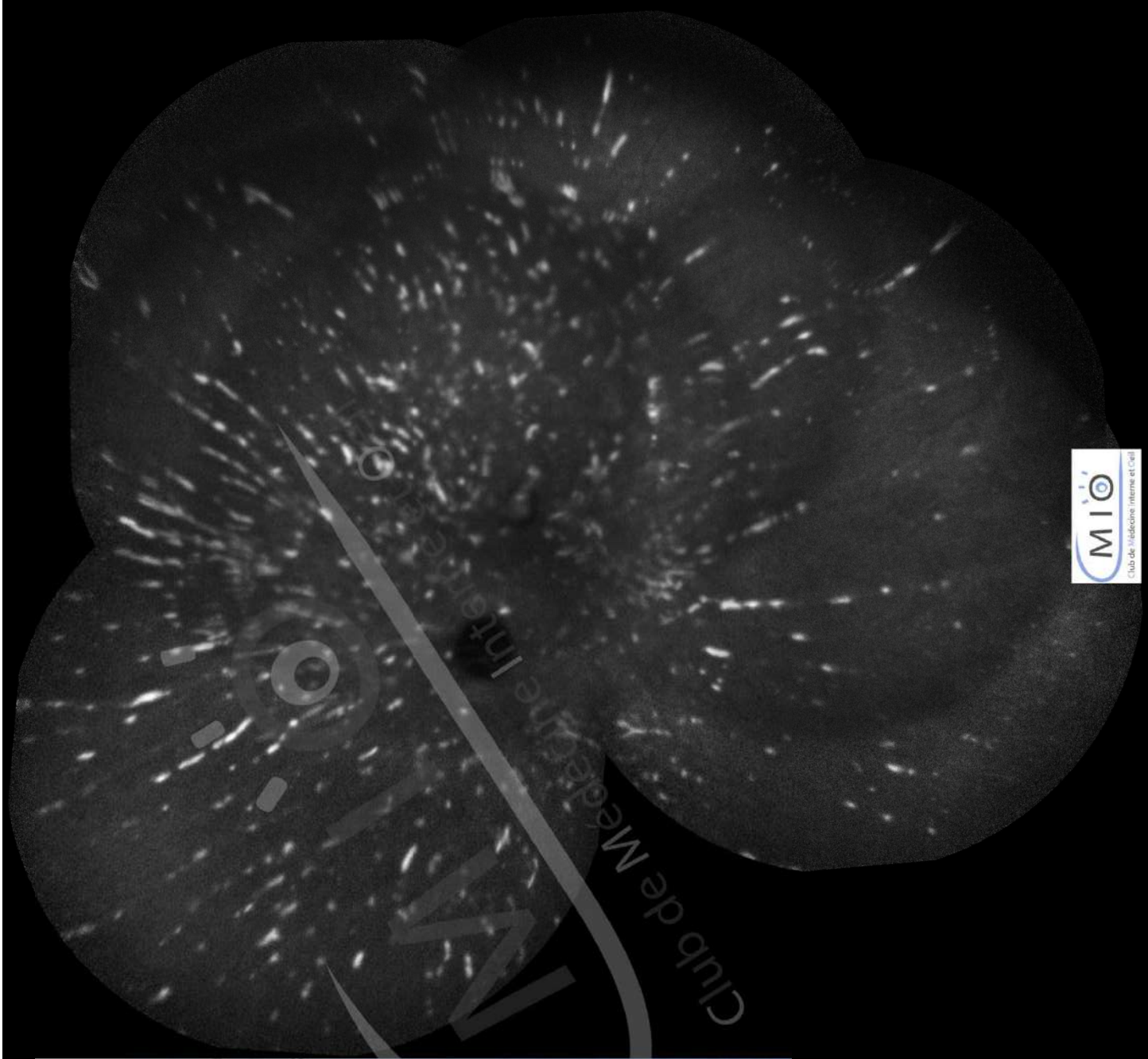
24 mn

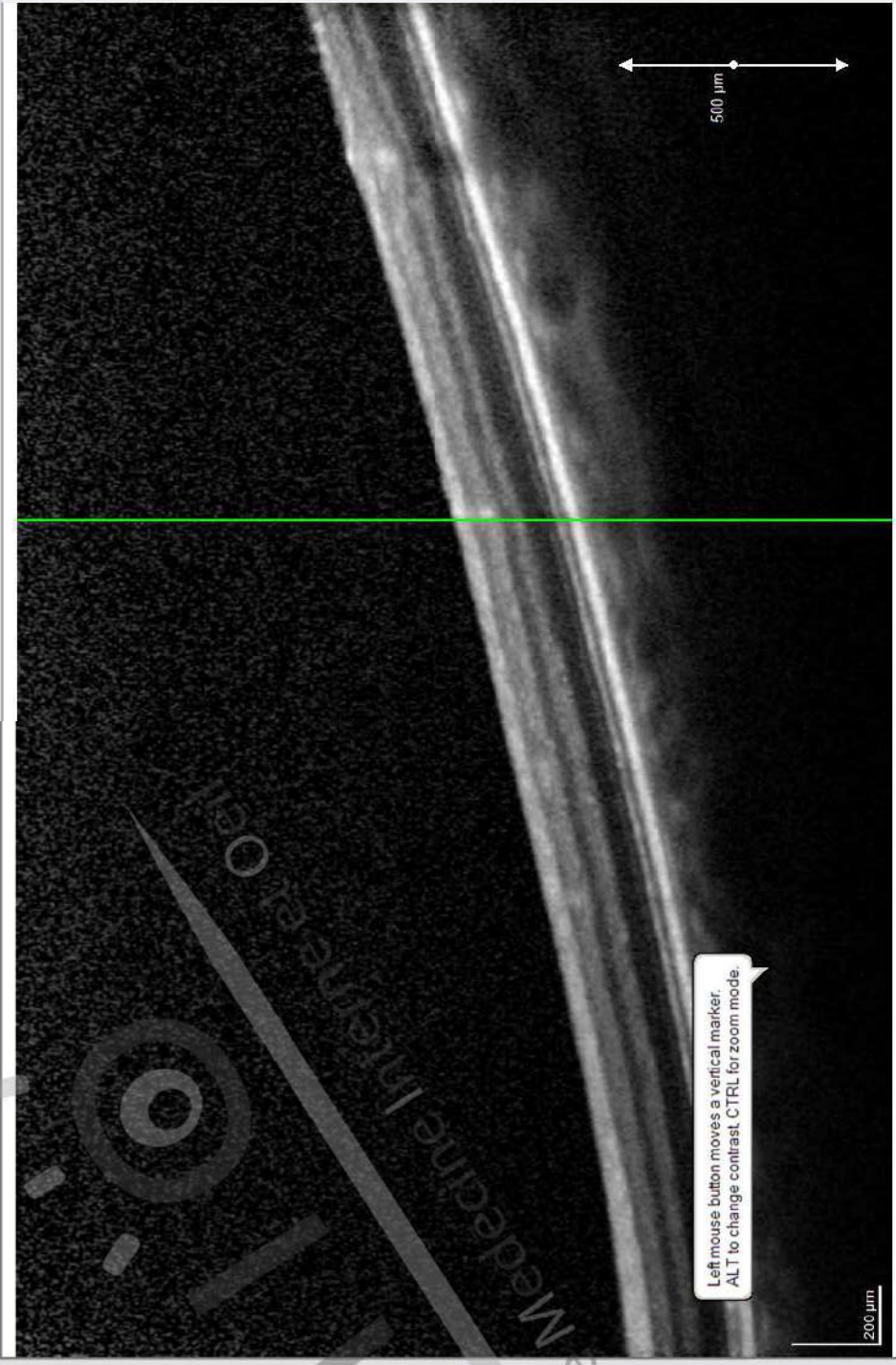
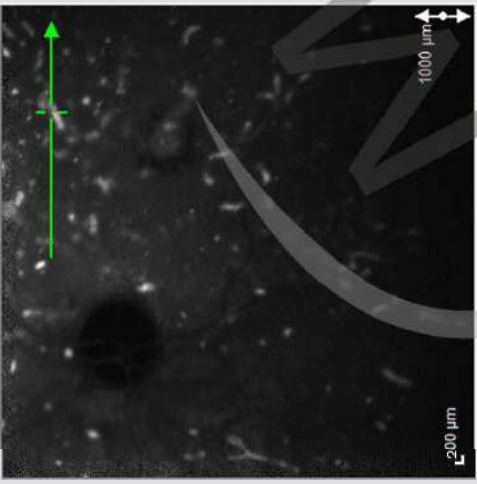




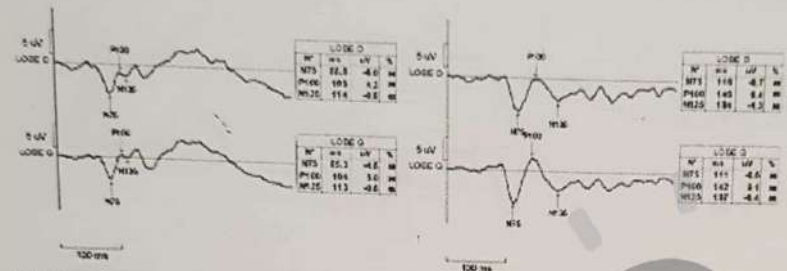




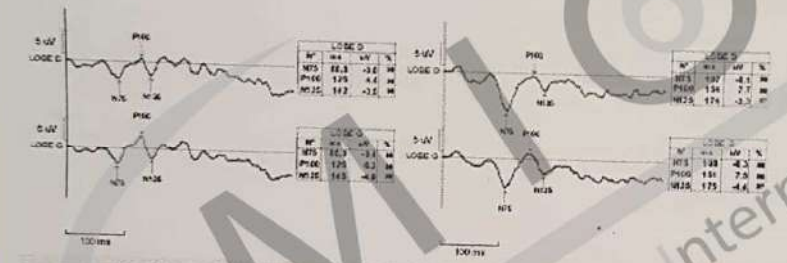




Electroretinogramme –
VEP : no abnormalitie



DAM-60
OG stimulé 1mn 0s Val= 51 Rej= 0
DAM-15
OG stimulé 1mn 46s Val= 31 Rej= 0

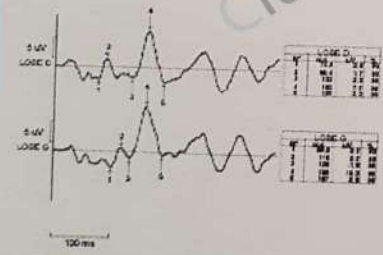


EXPLORATIONS FONCTIONNELLES
DE LA VISION MEB CHRU LILLE

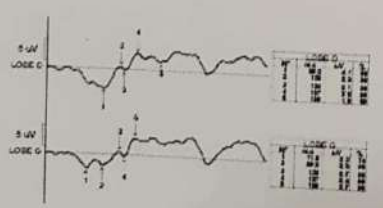
Operateur: Dorine BASSET
nom : GELEZ/BRETON/Raymonde
dossier : 10001520
date naiss : 12/11/1934

EXAMEN D'ELECTROPHYSIOLOGIE VISUELLE

PEVBLANC
OD stimulé 1mn 59s Val= 42 Rej= 0



PEVBLANC
OG stimulé 1mn 54s Val= 65 Rej= 0



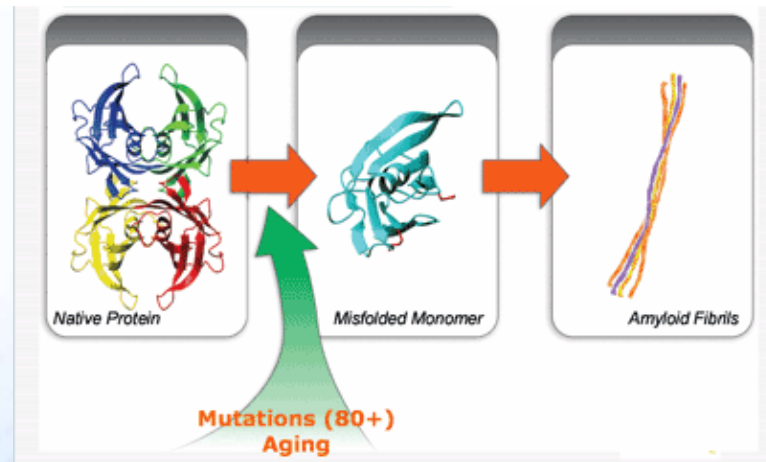


Medical history

- 2005 Difficulty of walking : Sensory-dominant Polyneuropathy
- 2007 : Admitted for anorexia, loss of weight, dysphagia, recurrent diarrhea and constipation
- Proteinuria : 1g/24h
- Syncopal episode
 - Autonomic nervous system assessment : cardiac frequency variability
- No CNS involvement

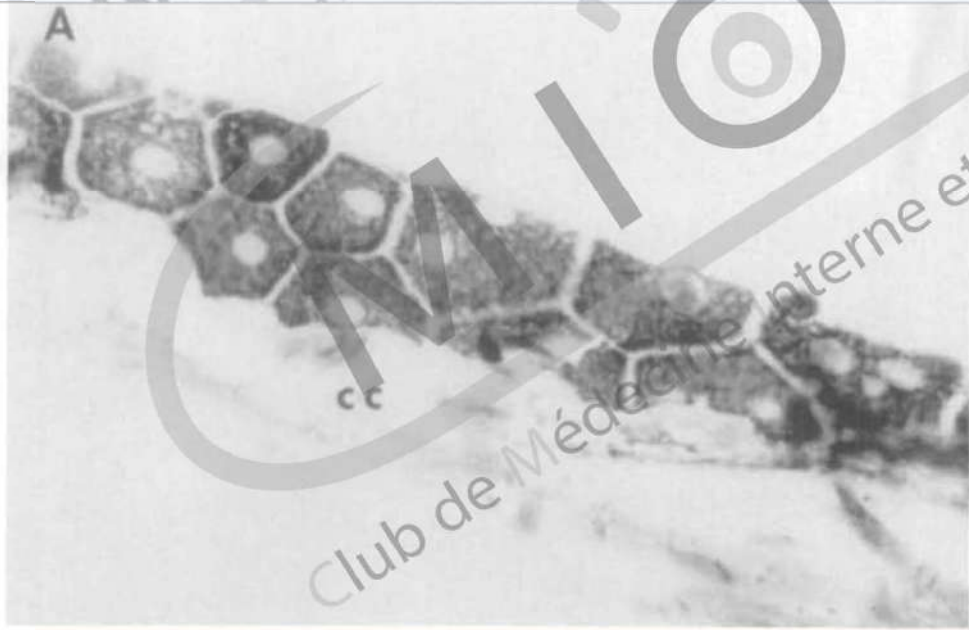
Medical history

- Familial amyloidotic polyneuropathy (FAP)
- 2007 : TTR gene analysis Mutation of transthyretine (Val30Met)
- Systemic disease ; polyneuropathy, cardiomyopathy, ocular involvement
- Autosomal-dominant diseases of variable penetrance caused by the deposition of polymerized mutated TTR in the peripheral nerves, gastrointestinal tract, heart, ocular tissues, and other organs
- To halt the progression of FAP : liver transplantation (was rejected in our patient)



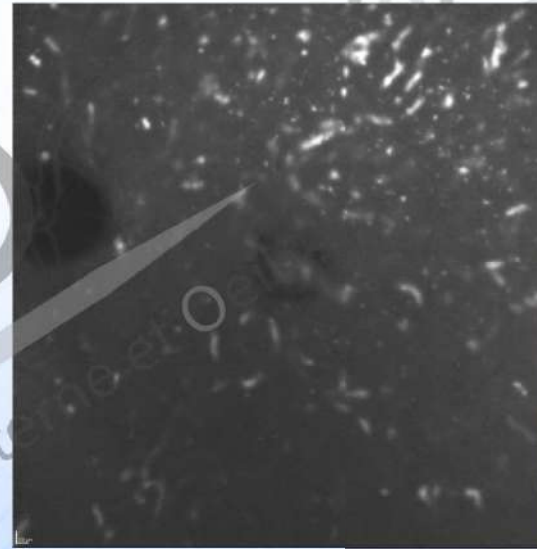
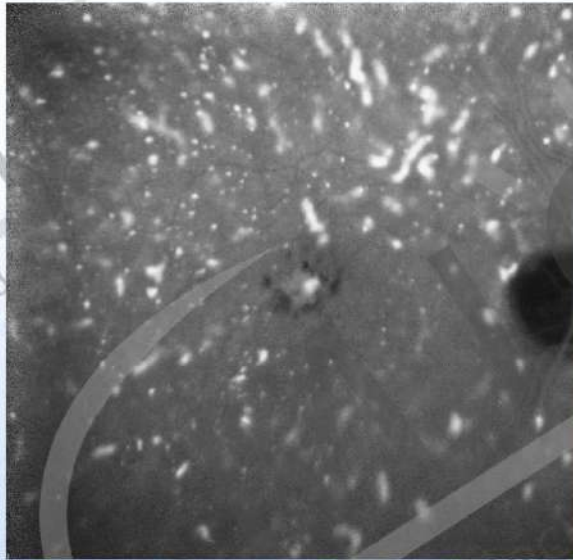
Distribution of Transthyretin in the Rat Eye

Andrew J. Dwork,*†‡ Tiziana Cavallaro,‡ Robert L. Marrone,‡ DeWitt S. Goodman,§
Eric A. Schon,‡‡ and Joseph Herbert*‡



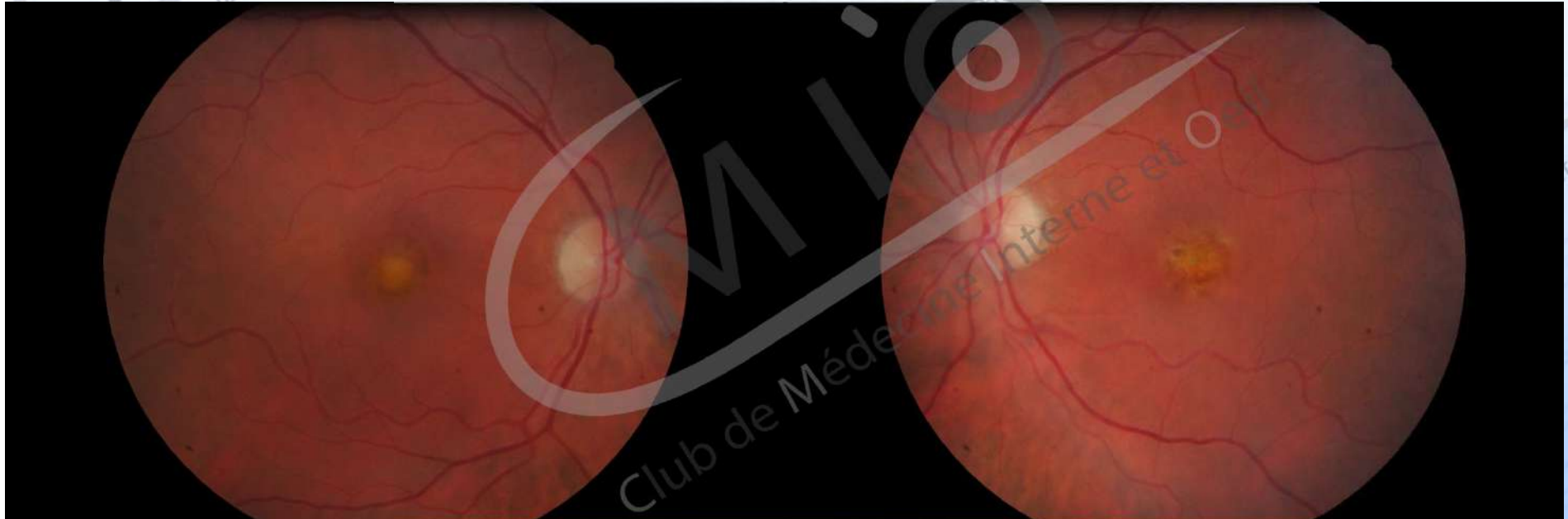
Intense immunoreactivity in the RPE

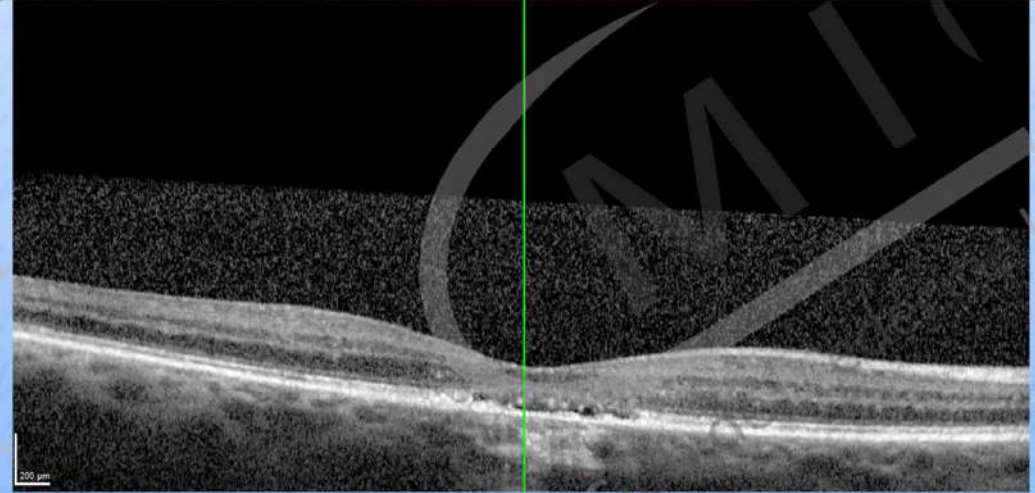
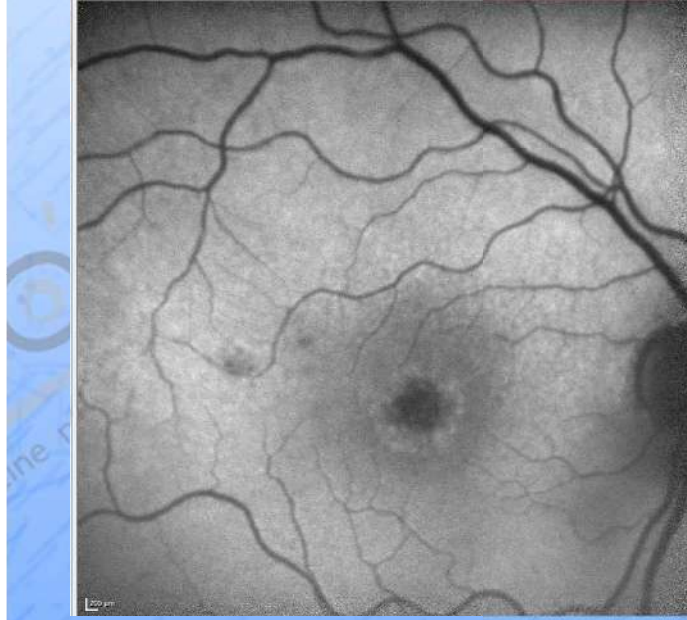
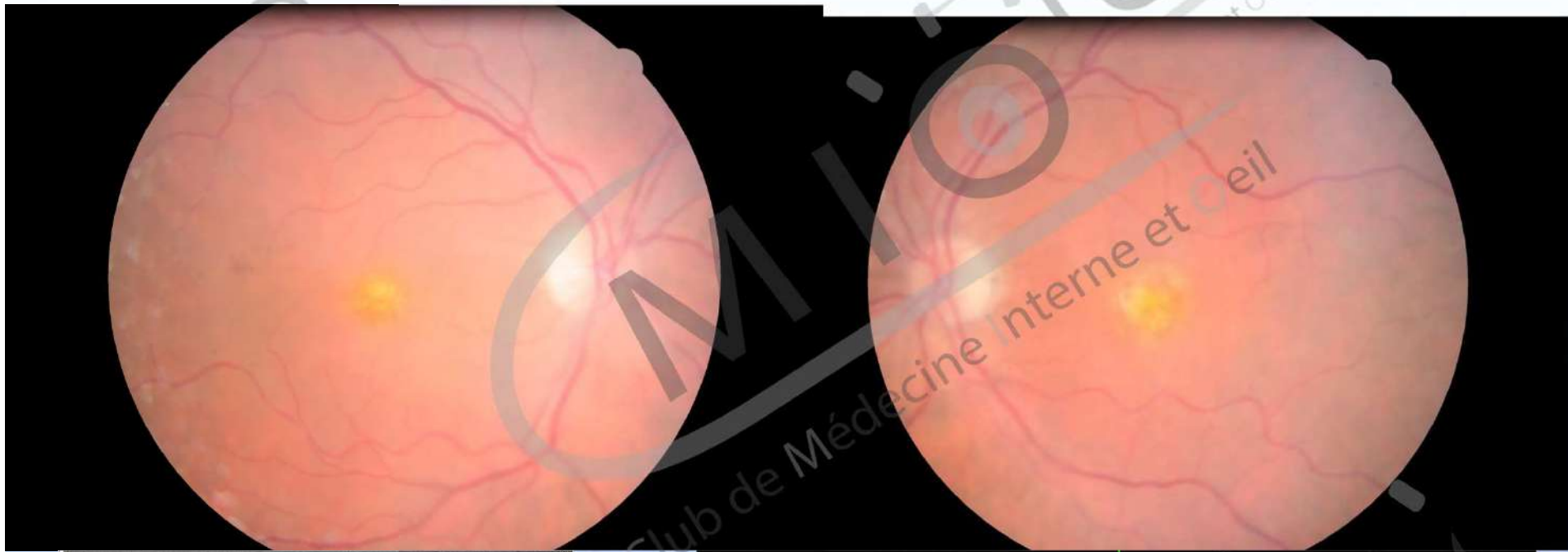
- Systemic :
Transthyretin mRNA is synthesized in the liver
- Eye : TTR is synthesized in the RPE and is transported in specific locations within the eye (choriocapillaris, retinal vessels, vitreous cavity)



- ICG dye diffuses through the choroidal stroma during angiography, accumulating within the RPE cells
- Binding of ICG dye to mutant transthyretine protein (amyloid fibrils) in the choriocapillaris stroma and the RPE

2012





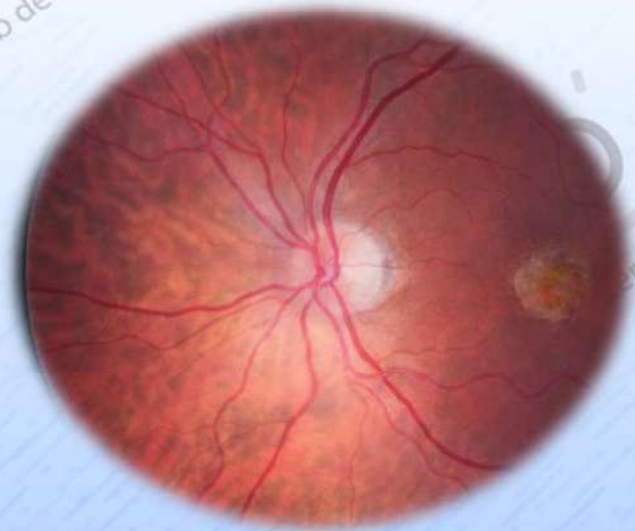


2017 : vitreous involvement of OD 9 years later of FAB
(Transthyretine-related familial amyloidotic polyneuropathy)

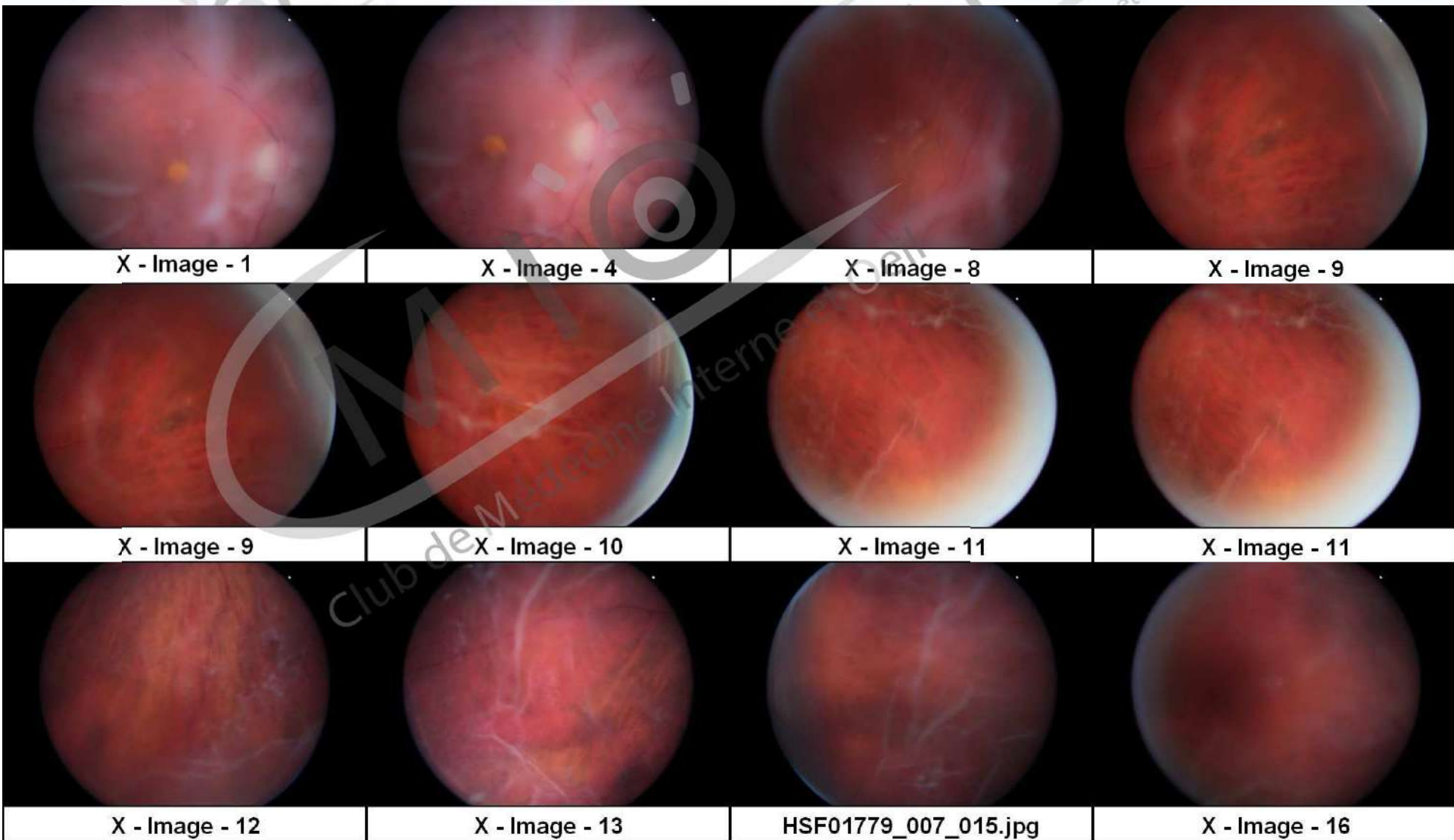
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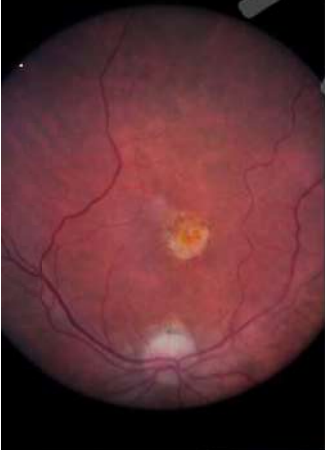


1,6/10

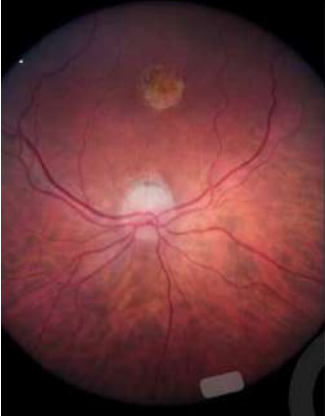


2017





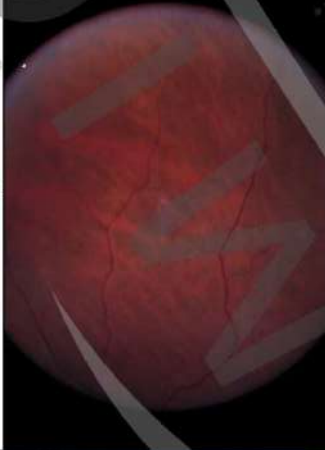
X - Image - 3



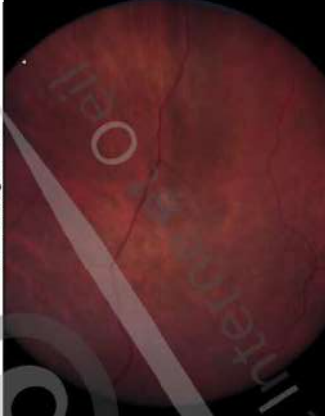
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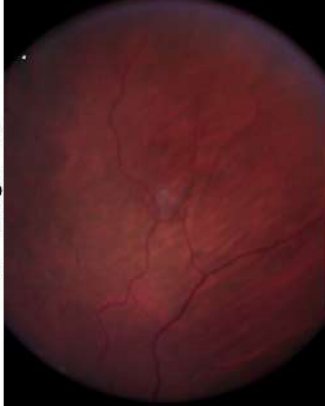
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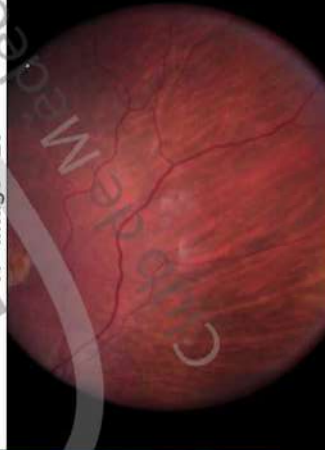
X - Image - 20



X - Image - 21



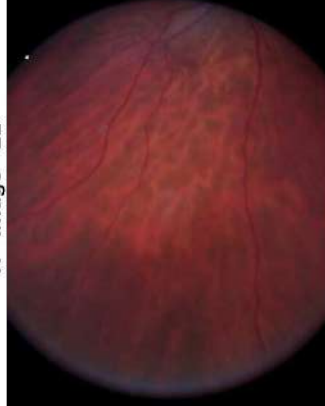
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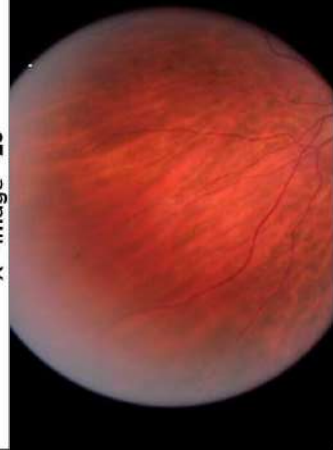
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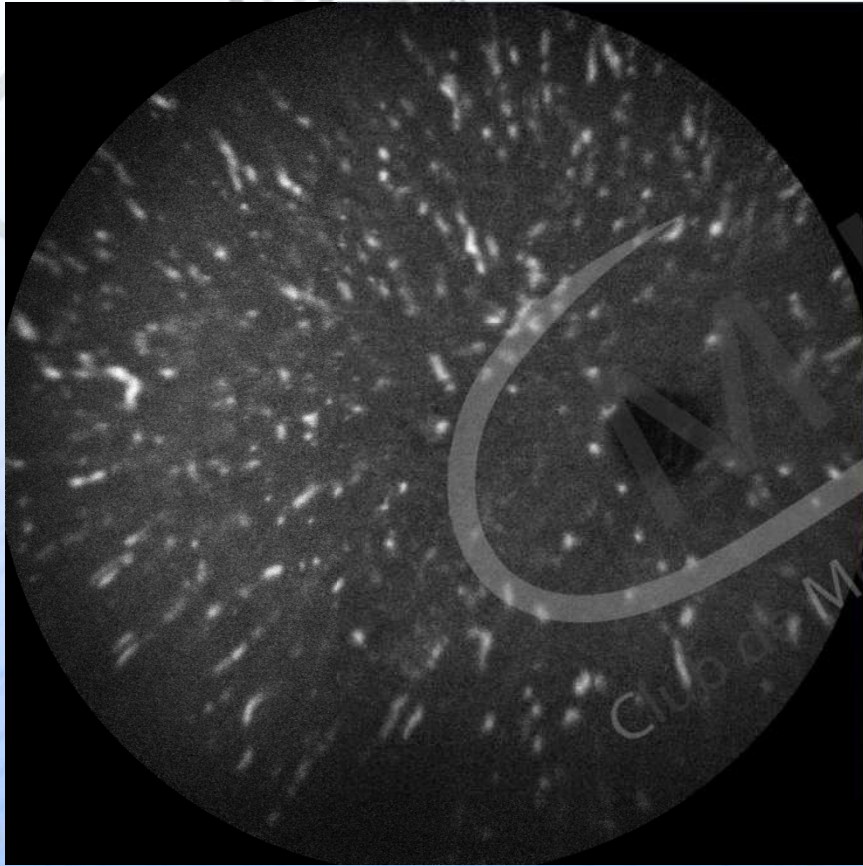
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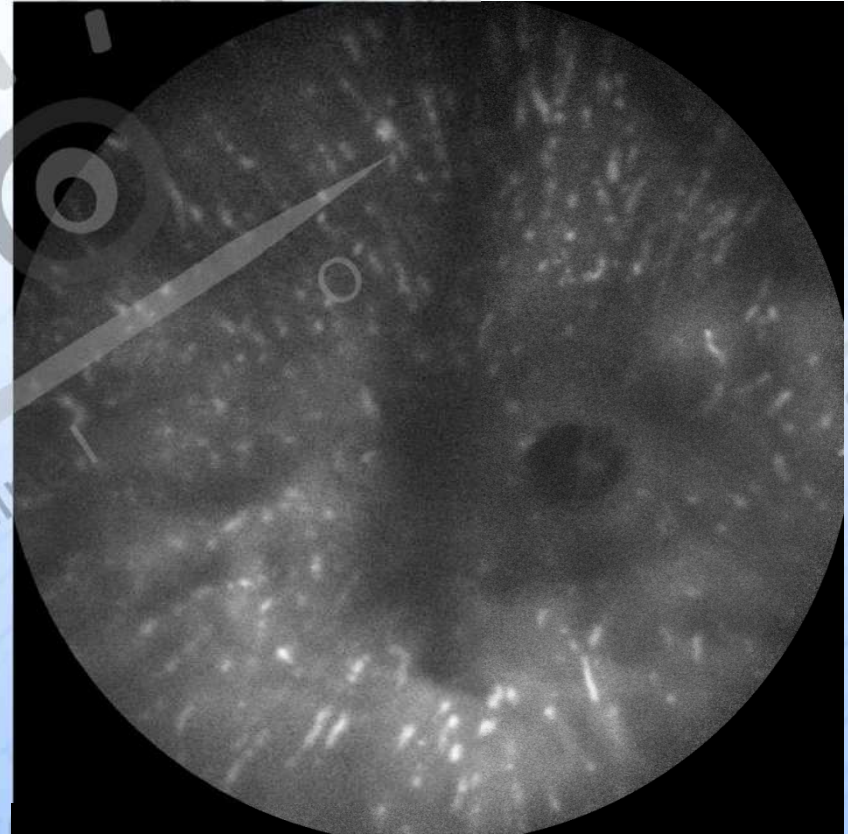
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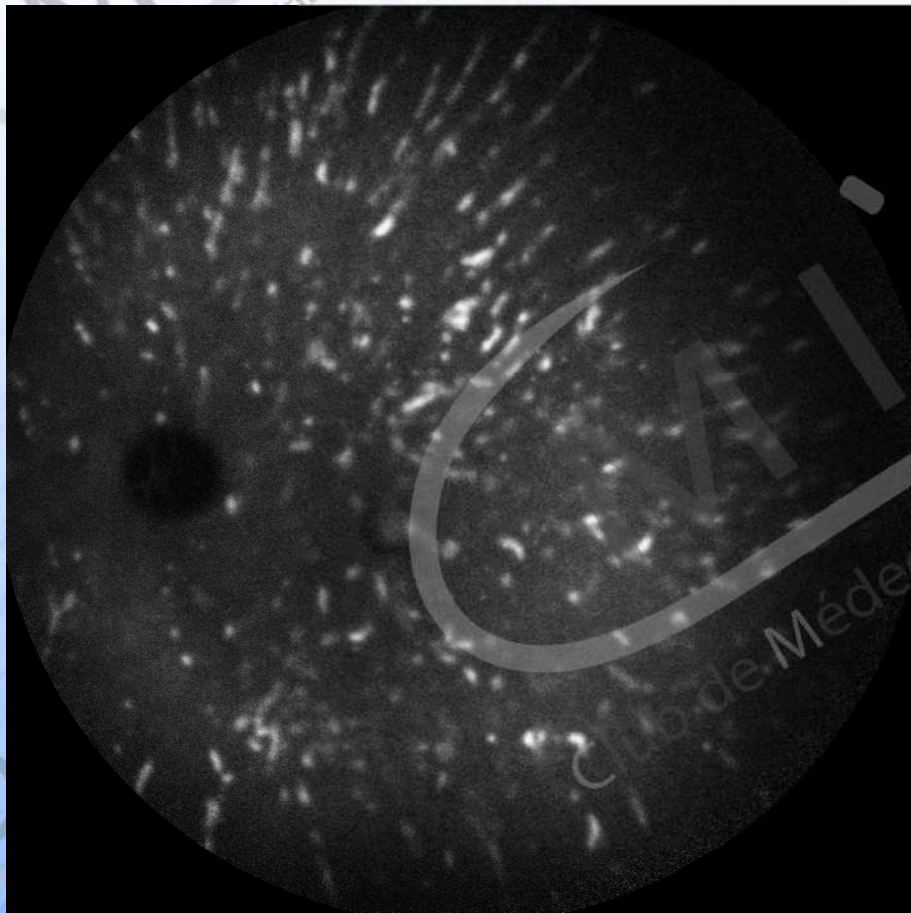
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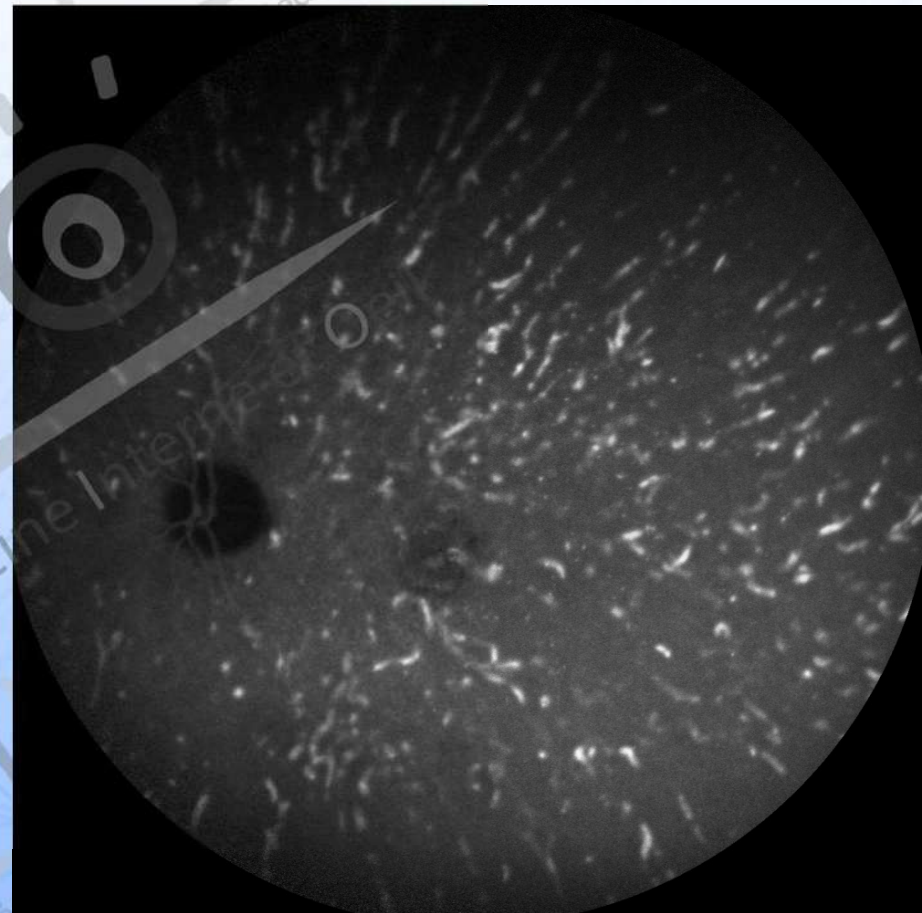
2010



2017

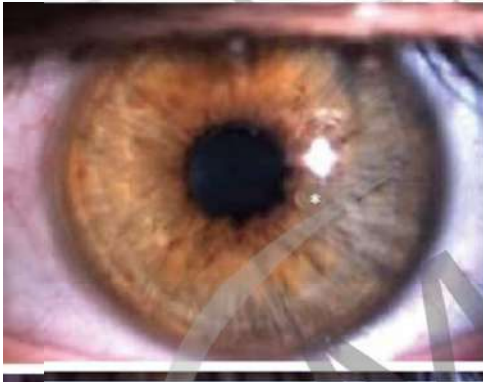


2010



2017

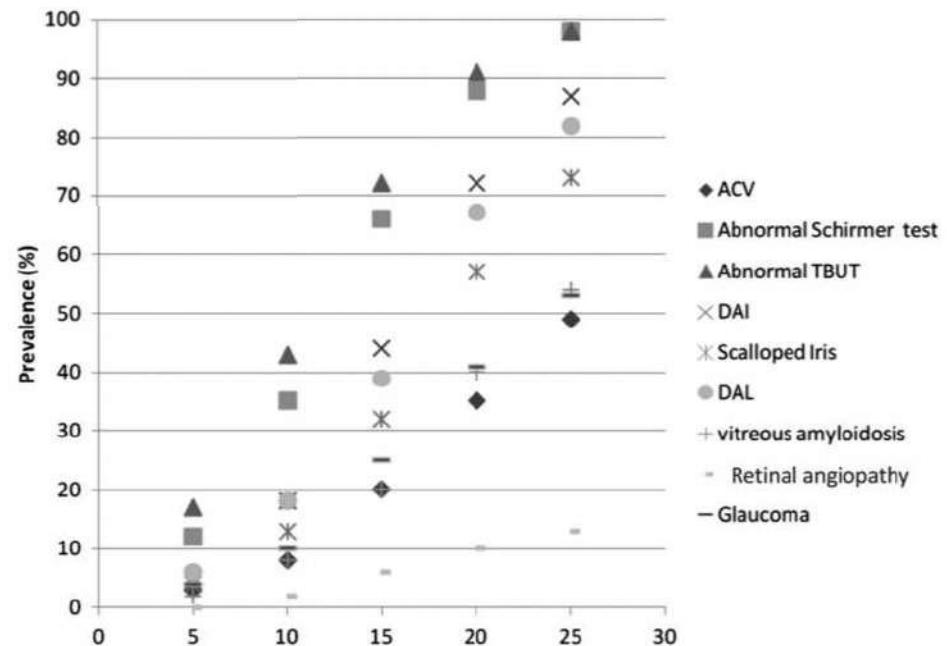
Ophthalmological manifestations of FAB



- Caused by deposition of amyloid in various intra-ocular tissues: Vitreous, iris, pupillary border, anterior capsule and trabecular meshwork.
- Progression of ocular amyloid deposition even after hepatic transplantation

120 J. M. Beirão et al.

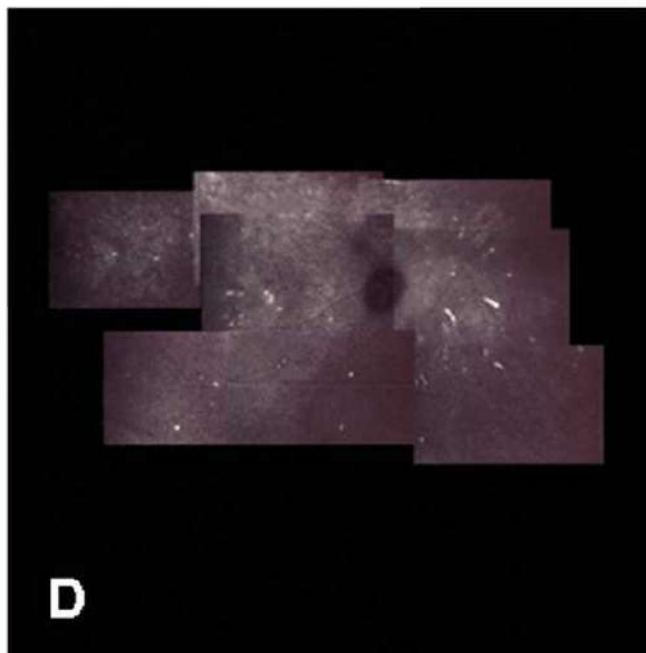
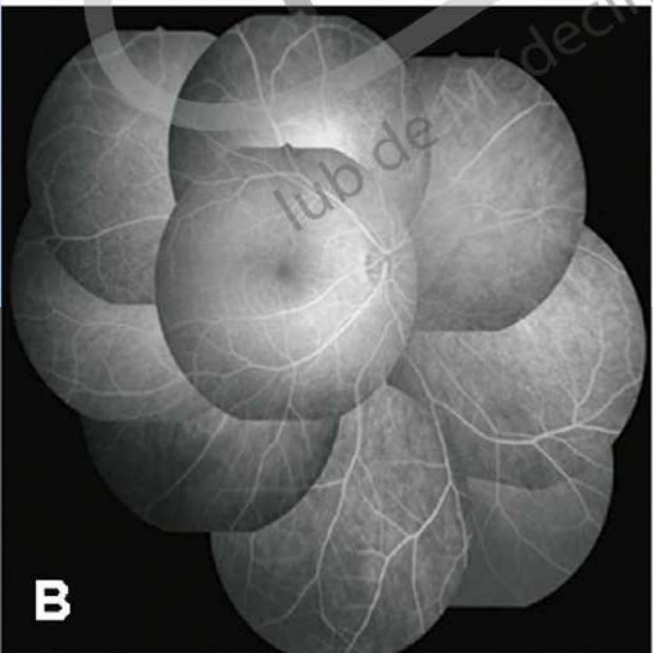
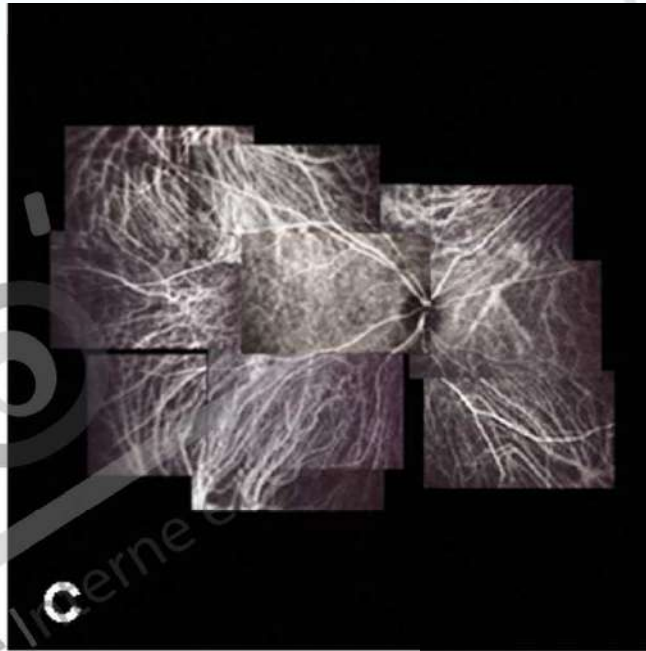
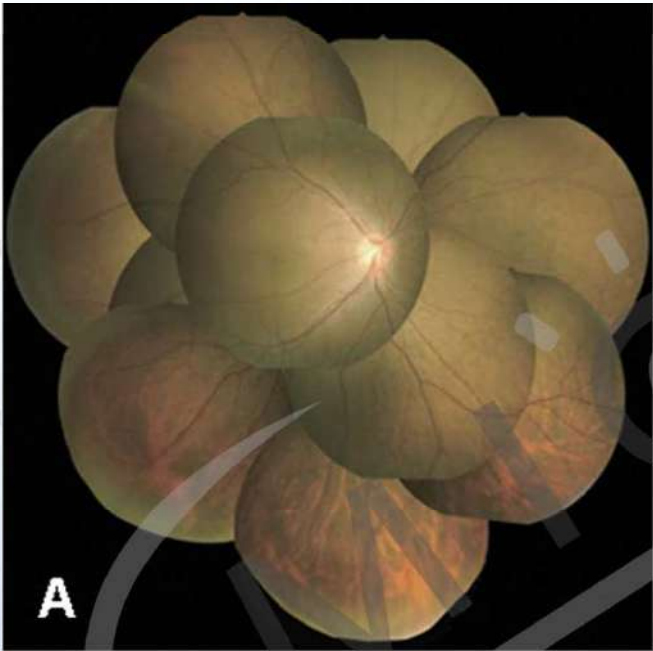
Figure 1. Prevalence of each ocular manifestation at 5, 10, 15, 20 and 25 years of disease. All prevalences increased with time. ACV, abnormal conjunctiva vessels; DAI, deposition of amyloid on the iris; DAL, deposition of amyloid on the lens.



[Amyloid. 2015;22\(2\):117-22. doi: 22.](#)

Ophthalmological manifestations of FAB

- Glaucoma
 - high rate of complications
 - decompression retinopathy (33%)
 - bleb encapsulation (48%)
 - (57%) needed additional surgery
- Vitreous amyloid
 - Late manifestation (60 months)
 - Vitrectomy improved VA
 - need surgery 2- 3 y later



MIÖ pattern
AB case
post
vitrectomy



[Kawaij,](#)
[Ophthalmology.](#) 200
5 Dec;112(12):2212

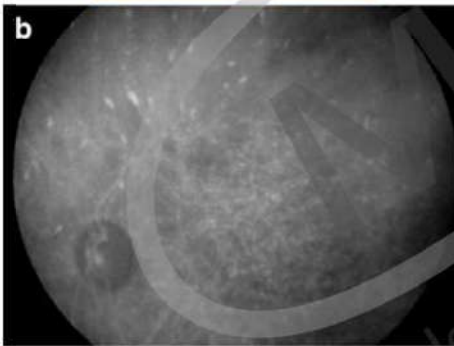
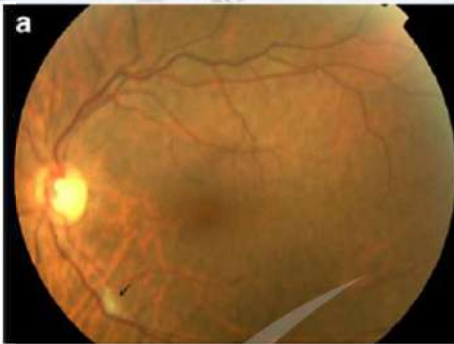
Same ICGA pattern in non familial amyloidosis

J Ophthalm Inflamm Infect
DOI 10.1007/s12348-012-0085-7

ORIGINAL RESEARCH

Indocyanine green angiography findings in patients with nonfamilial amyloidosis

Sonia Attia · Rim Kahloun · Sameh Mbarek ·
Oifa Harazallah · Habib Skhiri · Salim Ben Yahia ·
Moncef Khairallah



- 7 patients
- ICGA shows abnormal findings in all eyes (100%) with renal amyloidosis
 - Diffuse choroidal vascular staining (100%)
 - Hyperfluorescent streaks that closely follow the course of the choroidal vessel
 - Visible at late phase
 - Predominating in the periphery of the retina
 - Multiple punctuate hyperfluorescences (71.4%)
 - Hypofluorescent area (85.7%)

Conclusion

- ICGA evidenced ocular amyloidosis at the early stage
 - amyloid fibrils binding to the dye
 - Subclinical
 - Typical choroidal involvement (amyloidosis choroidopathy)
- Consider ICGA in the subclinical of amyloidosis, beyond the clinical examination
- Further study is needed



Enjoy with our
colleagues in
systemic
disease !

