

# FUNDOSCOPY, A USEFUL TOOL FOR EVALUATION OF PATIENTS WITH AUTOIMMUNE DISEASE

A. Stavrakakis, Consultant Eye Surgeon, Department of Retina ,  
University Hospital of Heraklion



*Fundoscopy* is primarily used to examine the vitreous, retina and blood vessels

## **WHEN & WHY**

- Rheumatologists often work in collaboration with ophthalmologists for a more comprehensive approach to patient care
- Certain autoimmune diseases are known to have ocular manifestations ( SLE, Behcet etc. )

and can be related with sight threatening complications

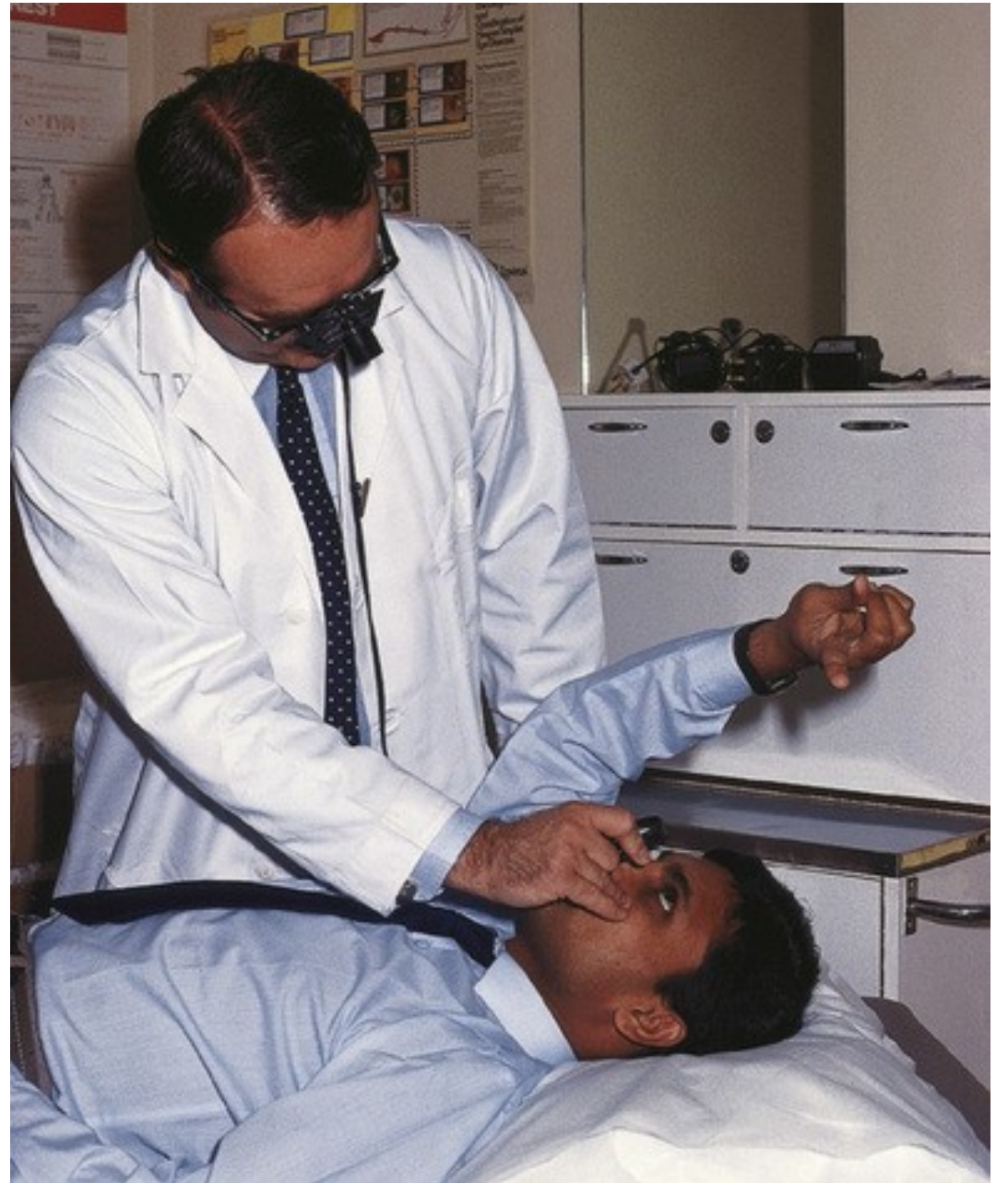
- Regular fundoscopic examinations may be recommended for
  - Cases of known or suspected eye involvement
  - Assessing effectiveness of treatment ( escalation if eye complications present )
  - Assessing possible toxicity of treatment ( Hydroxychloroquine )

Examination of the structures of the fundus using an ophthalmoscope can reveal disease of the eye itself or may reveal an abnormality indicative of disease elsewhere in the body

-Direct ( pupil dilation not necessary )



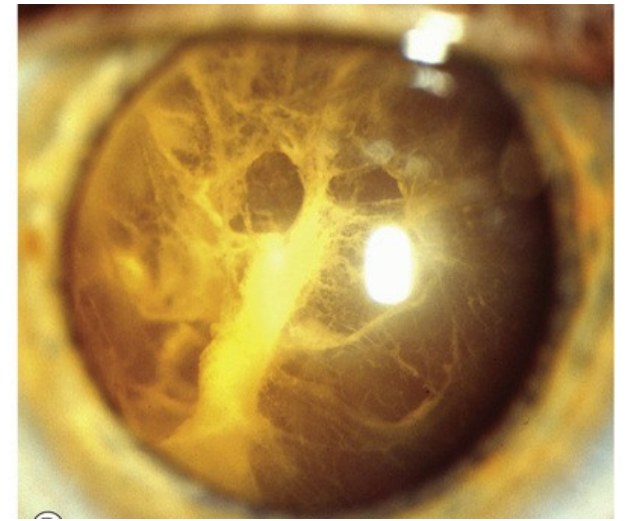
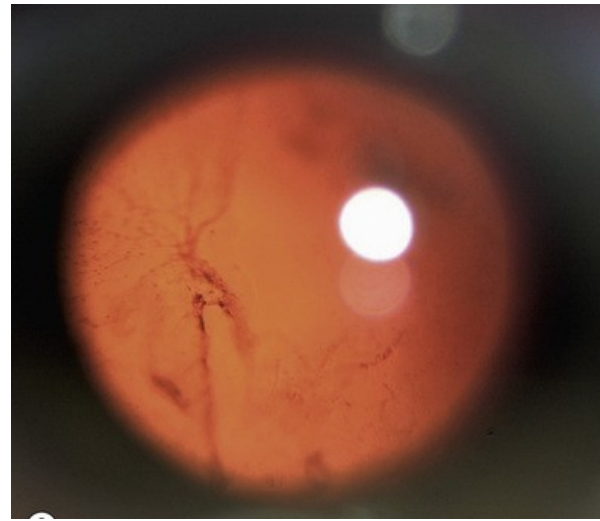
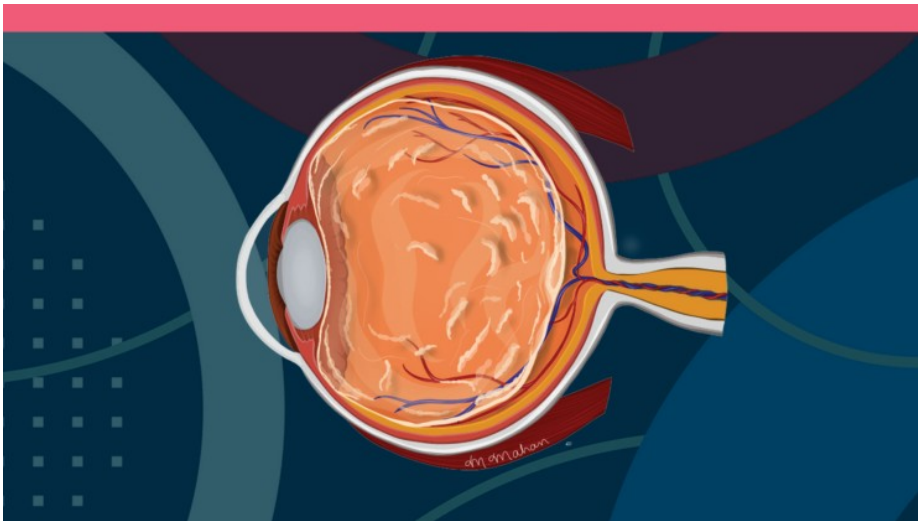
-Indirect ( pupil dilation needed )



Structures examined with fundoscopy :

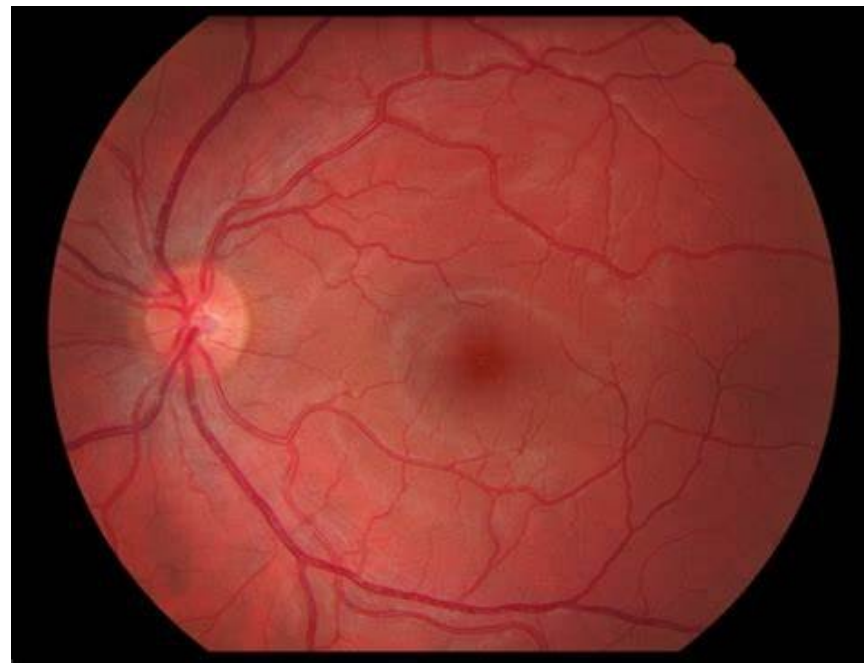
**VITREOUS** - transparent extracellular gel consisting of collagen, soluble proteins, hyaluronic acid and water. The total vitreous volume is approximately 4.0 ml

The vitreous provides structural support to the globe while allowing a clear and optically uniform path to the retina



## POSTERIOR POLE

Optic disc

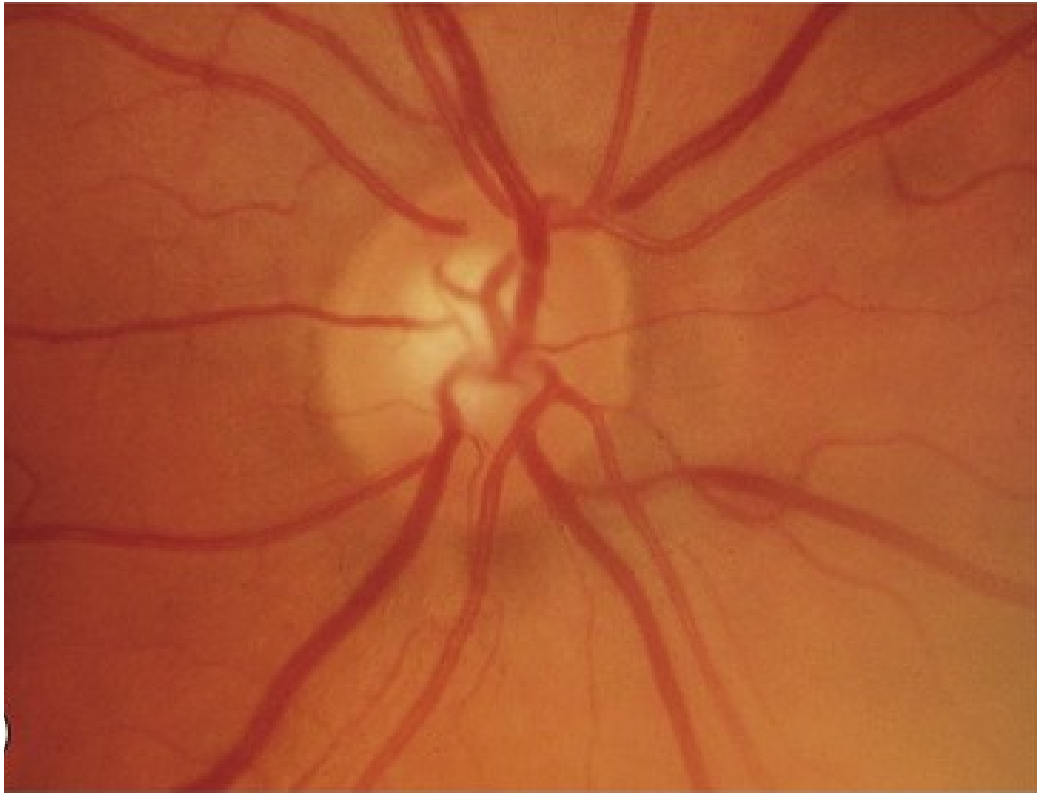


Major branches of the central retinal artery that emanate from the disc

Major branches of the central retinal vein collect at the disc

Macula-temporal to the disc-which appears darker ; no blood vessels are present in the center

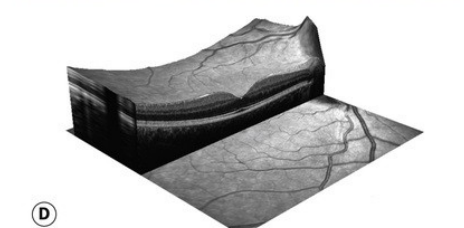
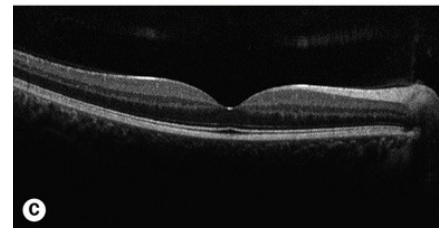
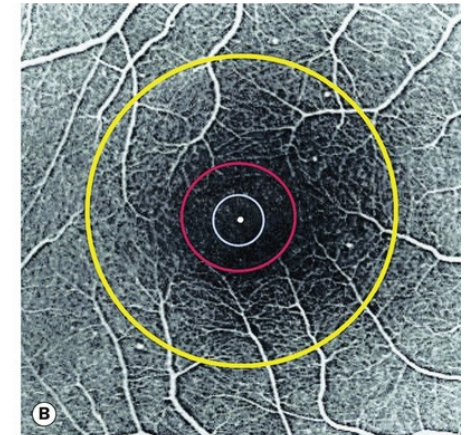
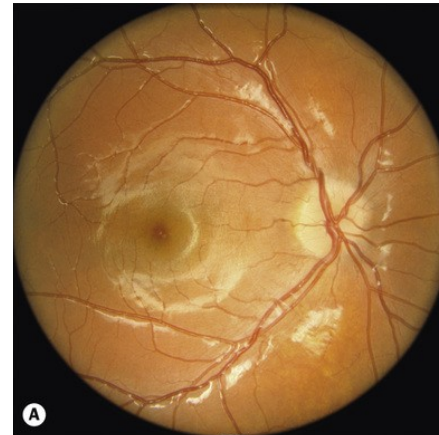
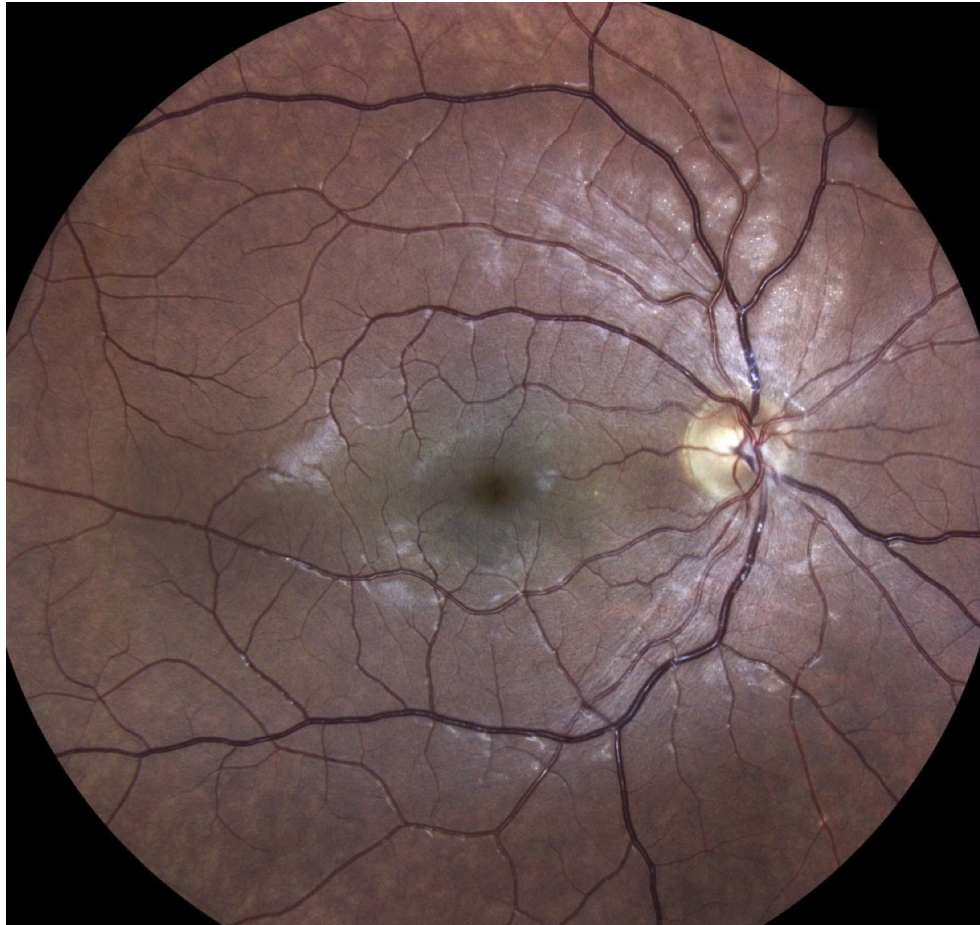
**OPTIC NERVE** –The intraocular segment (optic nerve head) is the shortest, being 1 mm deep and approximately 1.5 mm in vertical diameter. The ophthalmoscopically visible portion is called the **optic disc**





**MACULA** - A round area at the posterior pole, lying inside the temporal vascular arcades

It measures between 5 and 6 mm in diameter and subserves the central 15–20° of the visual field



# RETINAL VESSELS

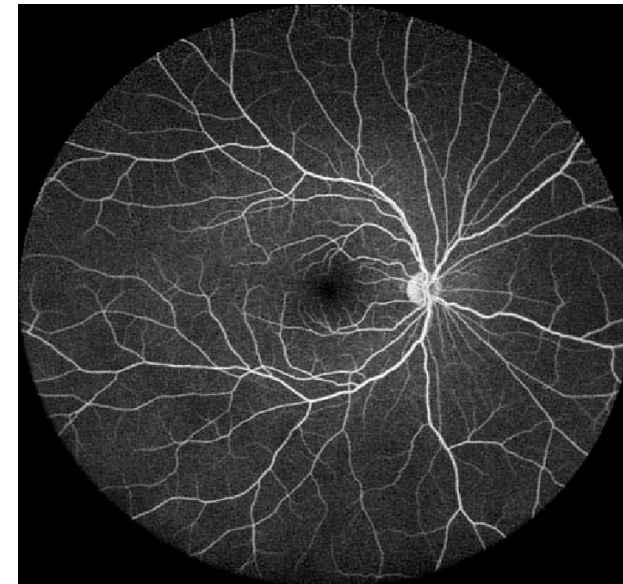
**Central retinal artery**, an end artery, enters the optic nerve approximately 1 cm behind the globe

**Retinal arterioles** that arise from the central retinal artery

## Venous system

Retinal venules and veins drain blood from the capillaries

Their diameter gradually enlarges as they pass posteriorly towards the **central retinal vein**

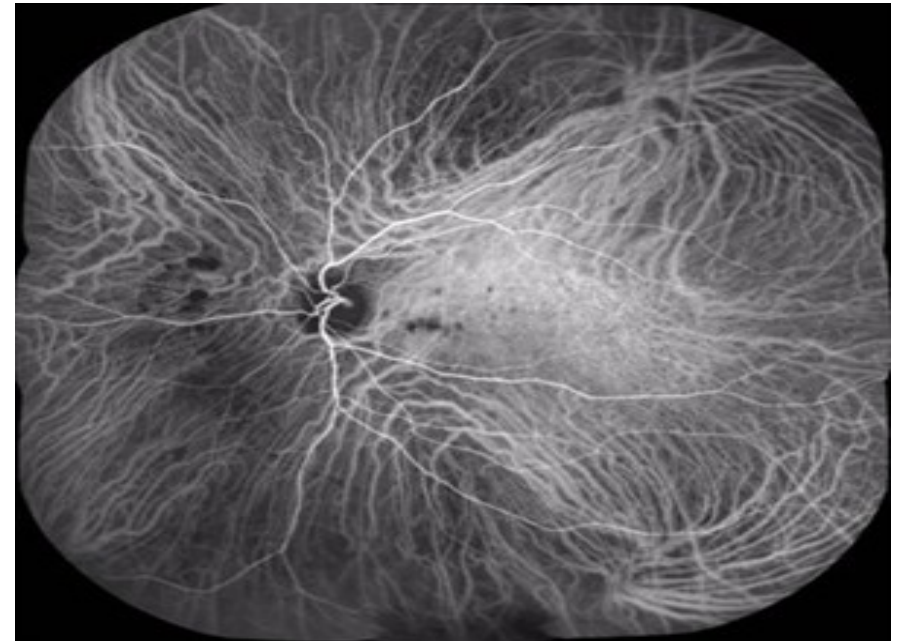
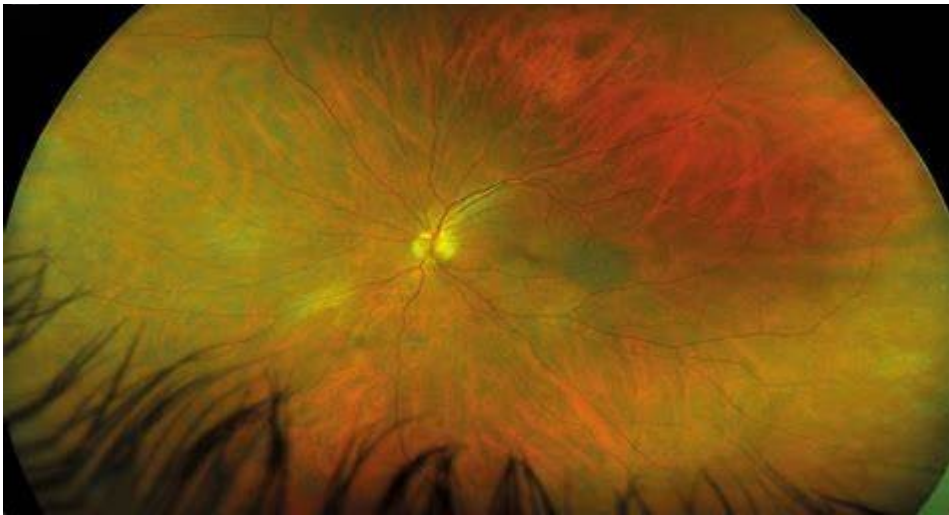


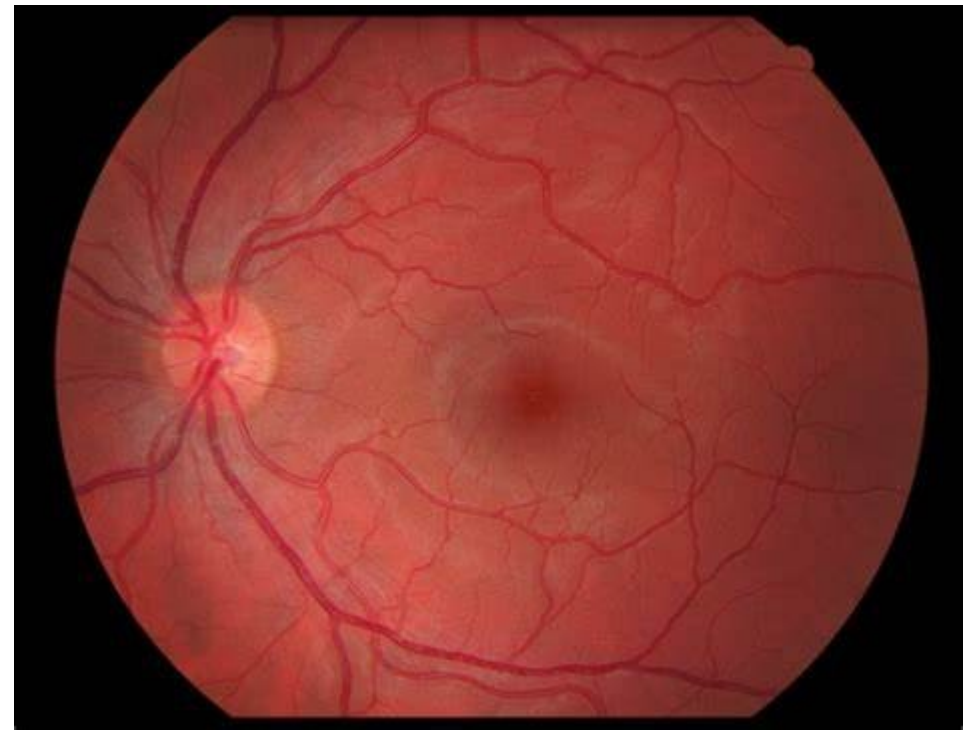
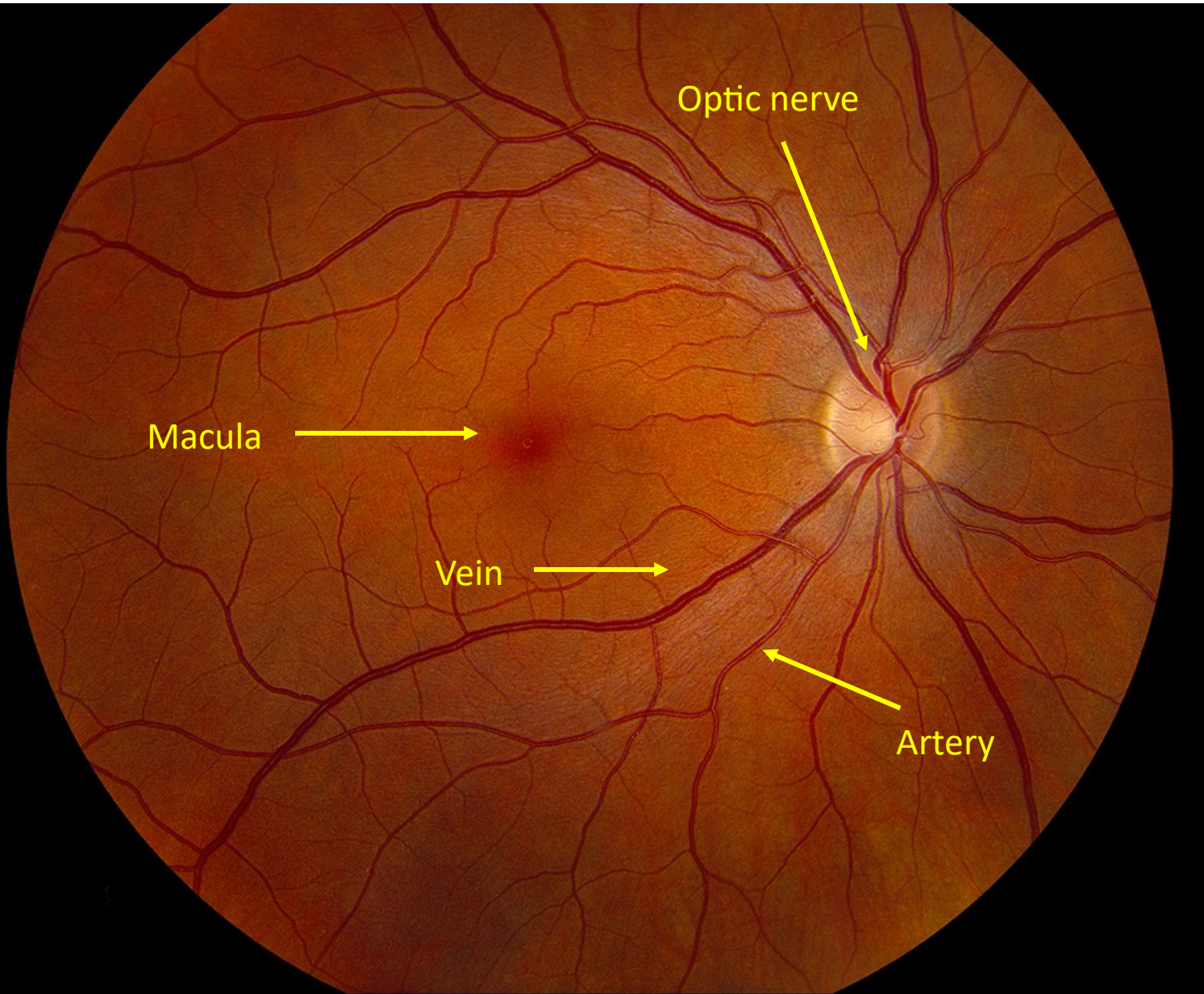
## CHOROID

The choroid is part of the uvea, and it contains blood vessels and connective tissue

The choroid is a highly vascular tissue, per unit weight, the choroid is the tissue with the highest blood flow in the body

Due to the vascular nature of the choroid, potentially any disease that affects systemic vasculature could potentially affect choroid health





Normal retina

A patient suspected for autoimmune disorder may have :

**VITRITIS** which may be asymptomatic or patient may complain for

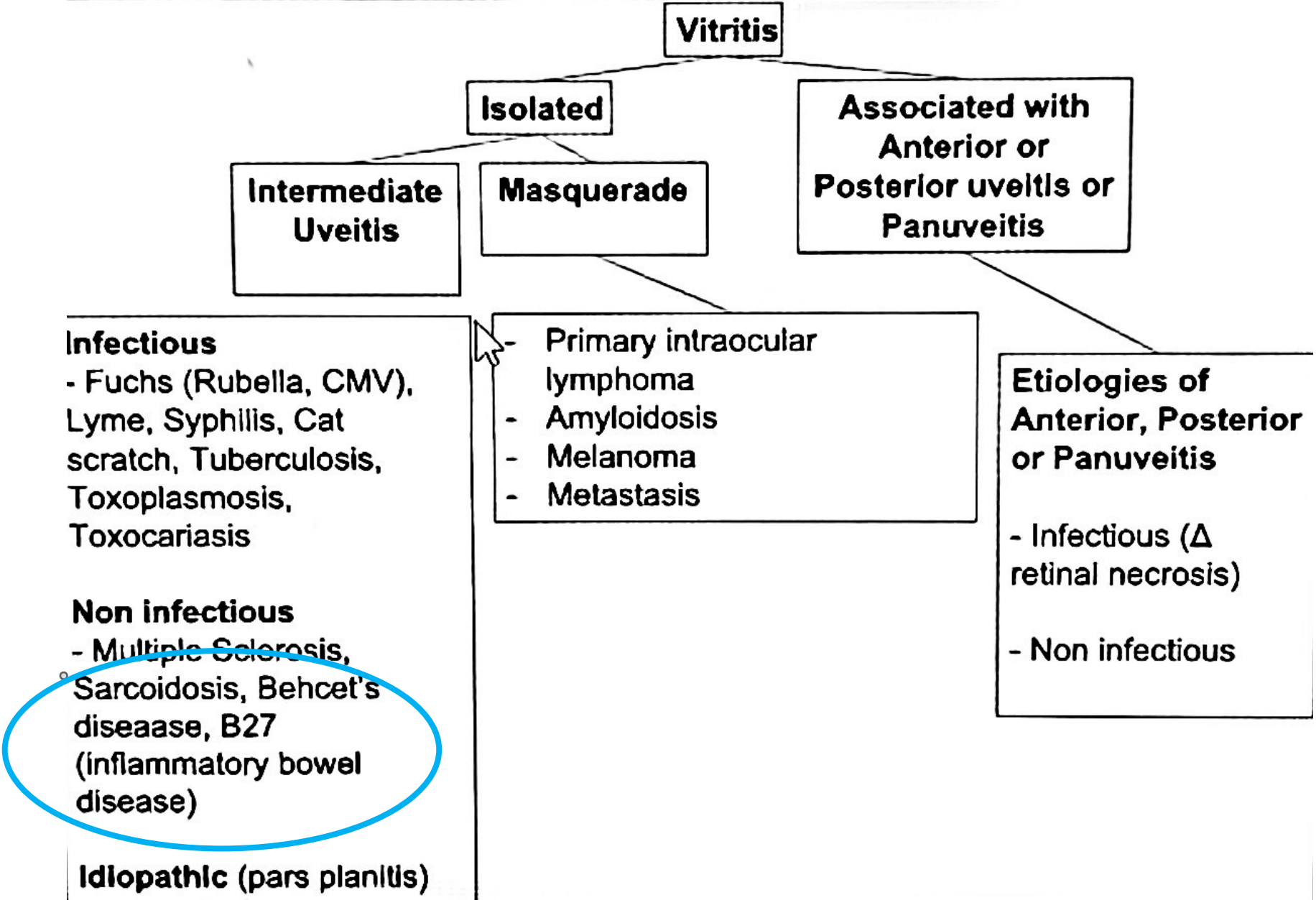
- floaters
- blurring of vision which may change with posture or eye movements
- reduced vision

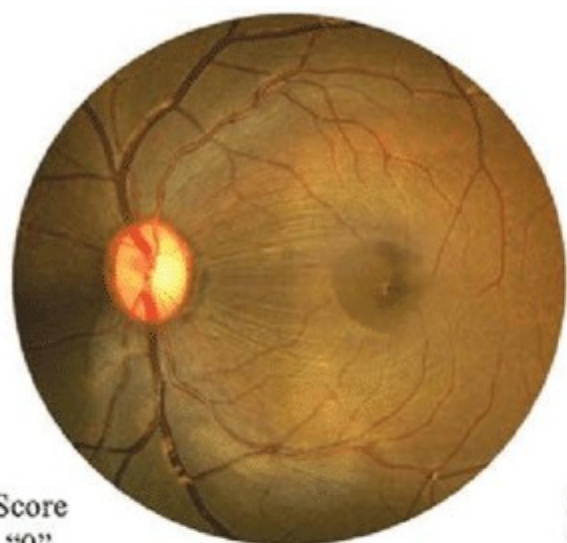
In intermediate uveitis , vitritis is the predominant site of inflammation

Cellular infiltration of the vitreous is causing haze and loss of transparency and is estimated with indirect ophthalmoscopy :

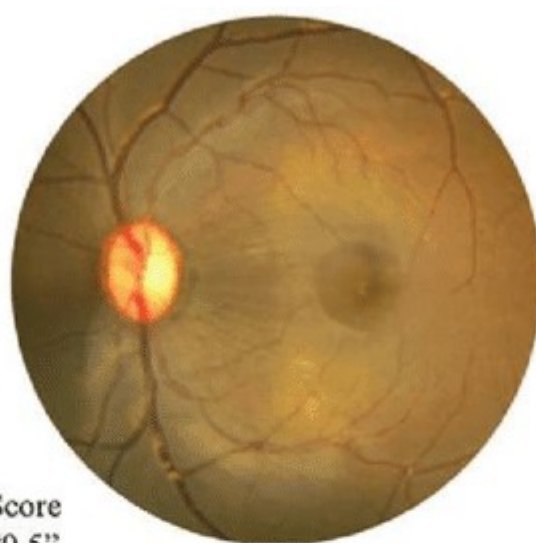
**Table 5: Grading of vitreous haze  
(Nussenblatt 1985 / National Eye Institute).**

Score	Description	Clinical findings
0	Nil	None
0.5+	Trace	
1	Minimal	Posterior pole clearly visible
2	Mild	Posterior pole details slightly hazy
3	Moderate	Posterior pole details very hazy
4	Marked	Posterior pole details barely visible
5	Severe	Fundal details not visible

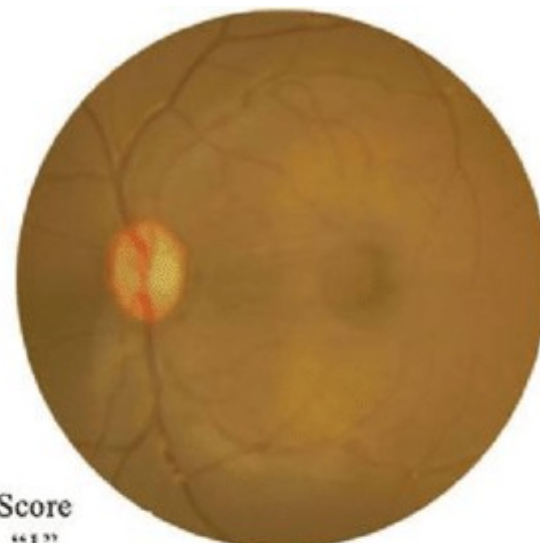




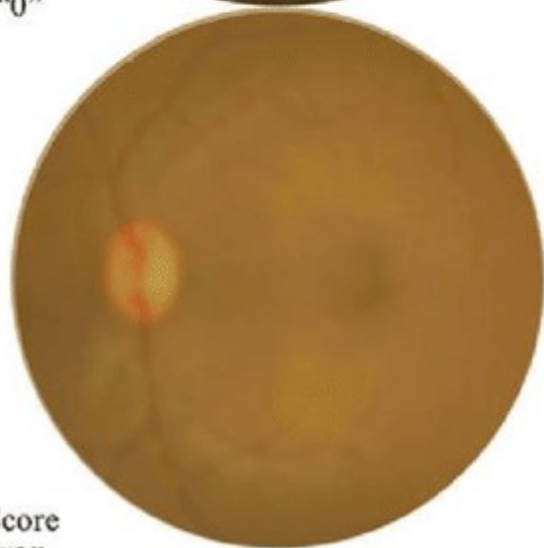
Score  
"0"



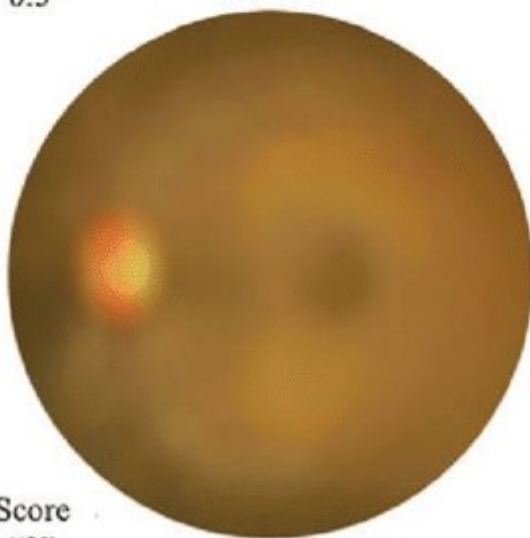
Score  
"0.5"



Score  
"1"



Score  
"2"



Score  
"3"



Score  
"4"



## OPTIC NERVE PATHOLOGICAL FINDINGS

- **Disk hyperemia**

Disk hyperemia may persist even in an eye with little clinically active inflammation elsewhere.

- **Papillitis**

- Demyelinating ( most common )

- Parainfectious ( post viral infection or immunization )

- Infectious ( sinus-related or associated with cat-scratch disease, syphilis, Lyme disease, cryptococcal meningitis and herpes zoster )

- Non-infectious ( sarcoidosis, systemic lupus erythematosus, polyarteritis nodosa and other vasculitides or optic neuropathies – arteritic ischaemic optic neuropathy )



- **Papilledema**

- bilateral swelling of the optic nerve head secondary to raised intracranial pressure (ICP)



- **Neovascularization** of the optic disc

- (a common finding in uveitis with retinal vasculitis and can regress with anti-inflammatory therapy )



- **Optic atrophy**

- may develop in the presence of ocular inflammation or after diffuse loss of retinal tissue

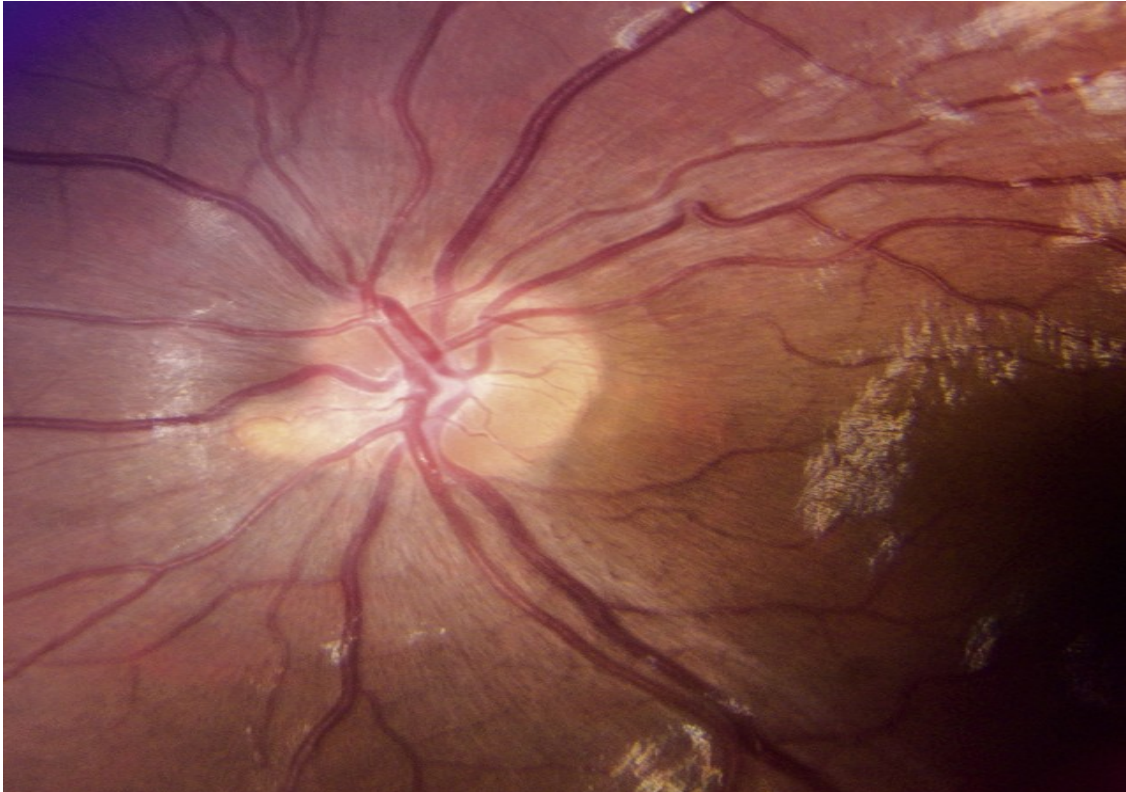
- flat white disc with clearly delineated margins

- reduction in the number of small blood vessels on the disc surface



- **Optic nerve granuloma**

- Granulomas may impinge on the optic nerve and optic disc in diseases, such as sarcoidosis

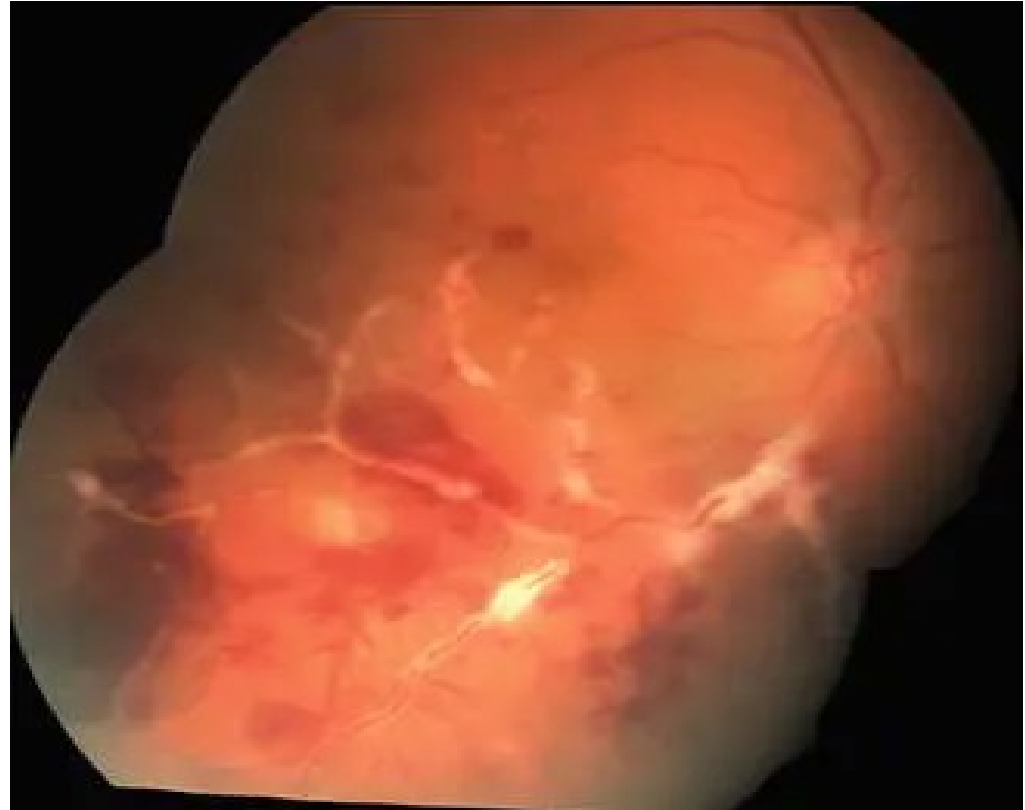
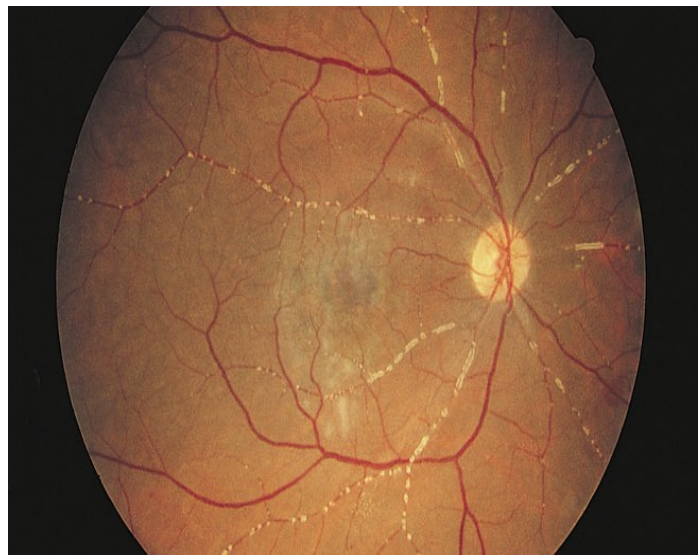


## RETINA & CHOROID

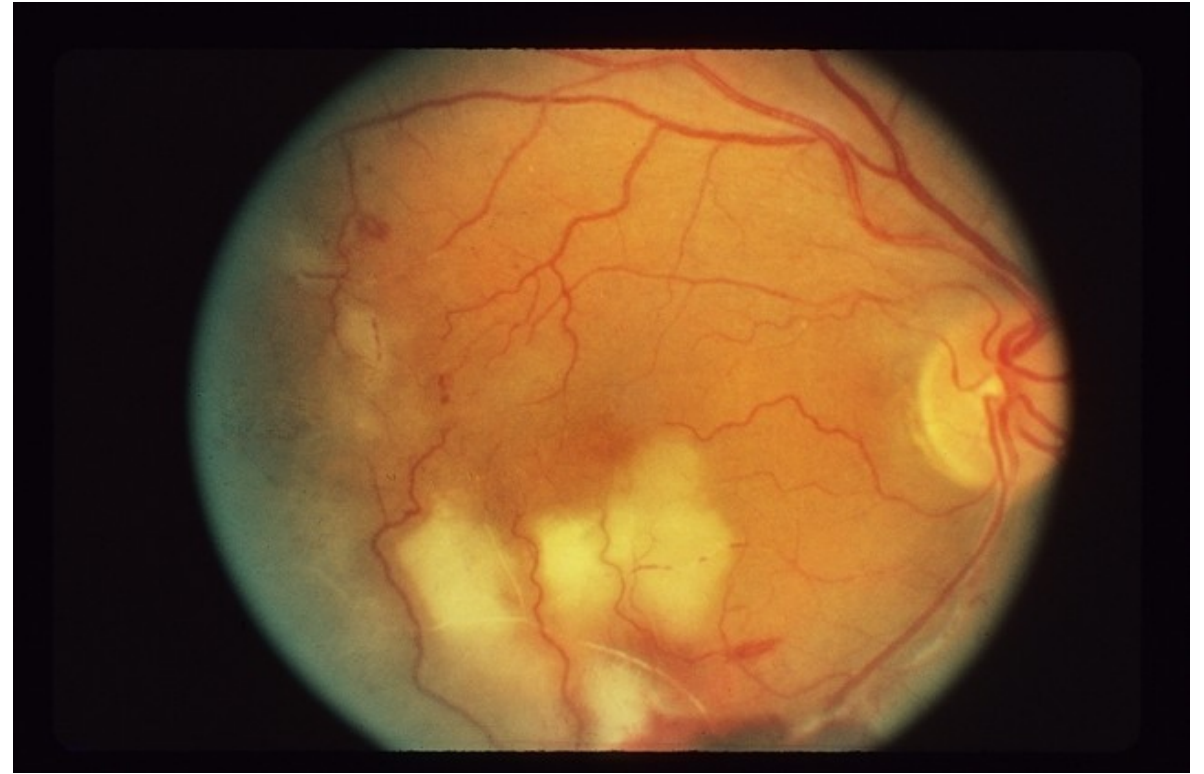
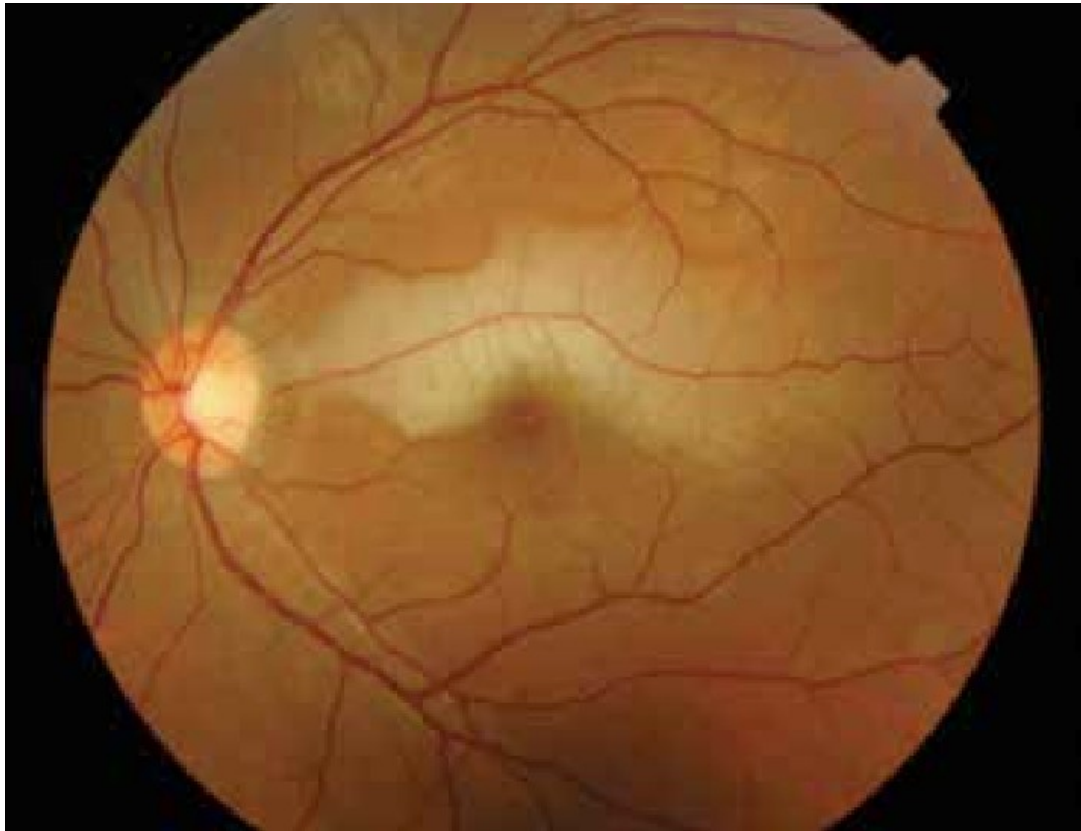
- Retinal vasculitis

- retinal vascular alterations are common in intermediate and posterior uveitis

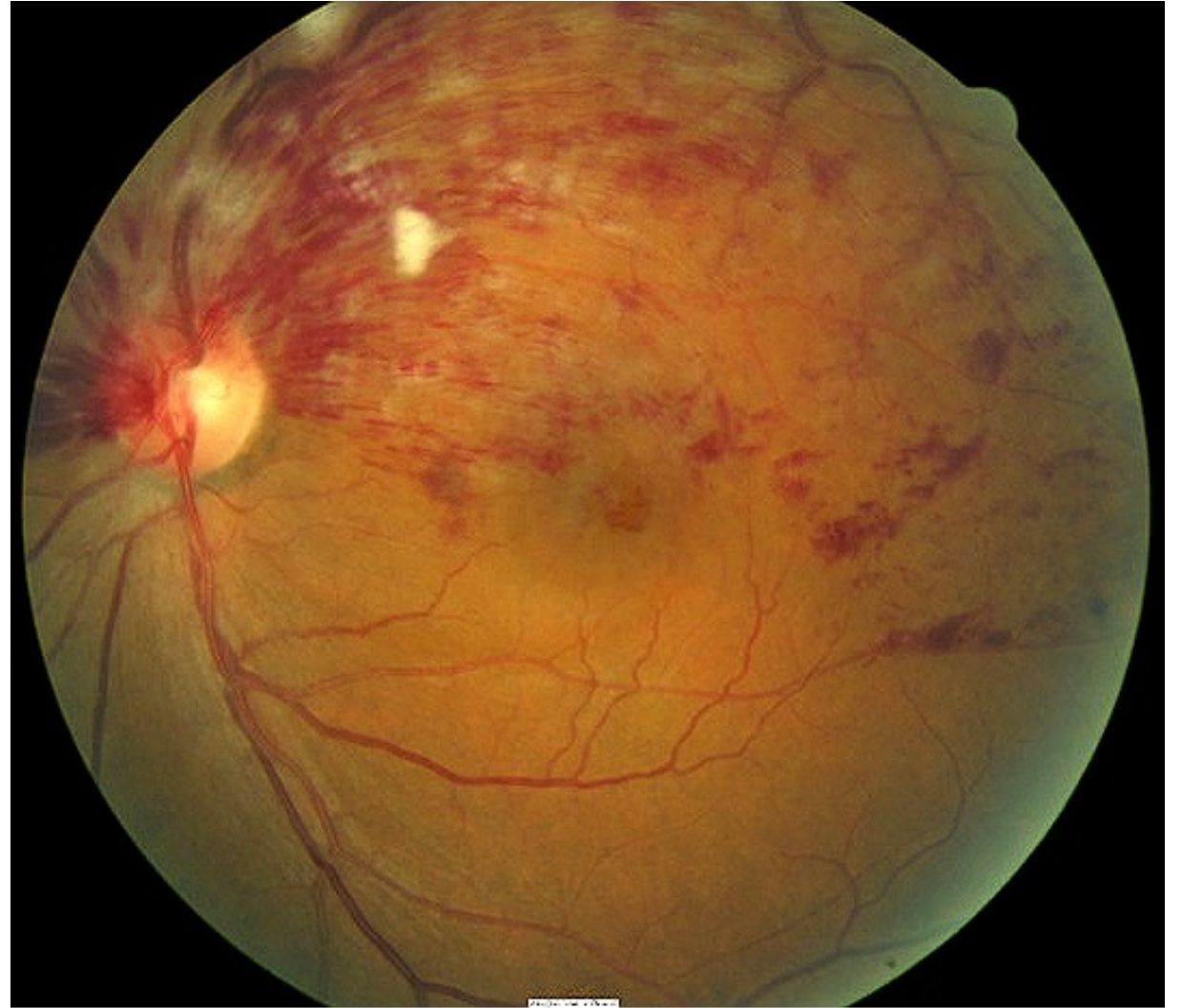
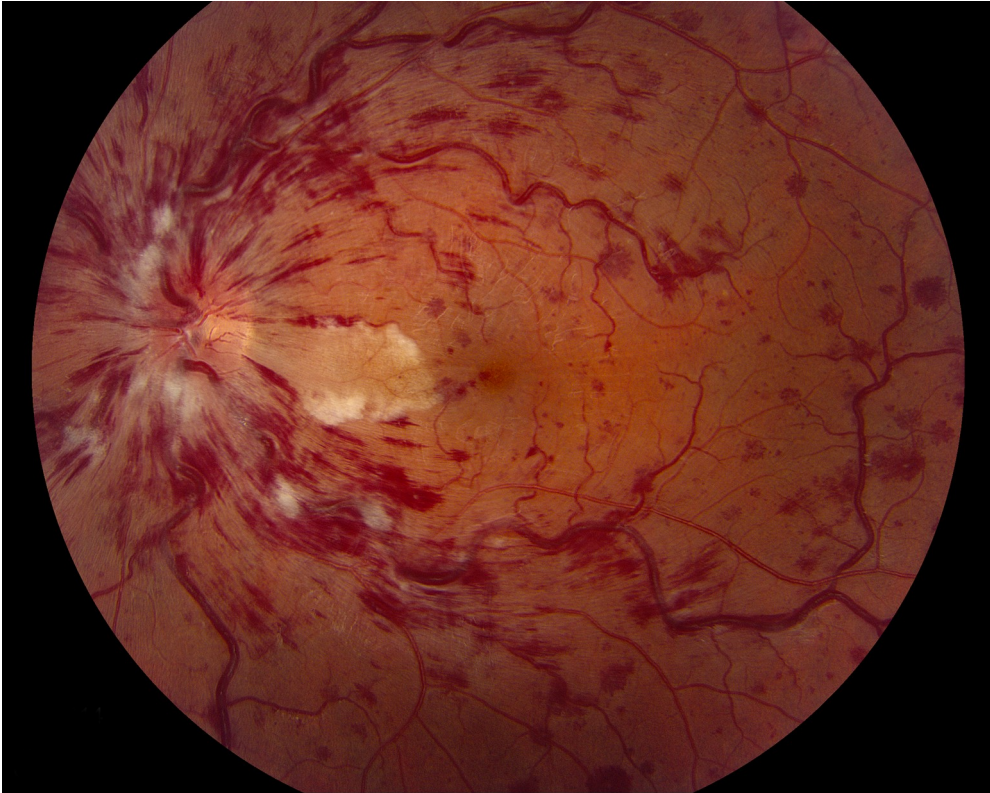
- vascular sheathing of the arteries or veins ( caused by infiltration of inflammatory cells around the vessels )

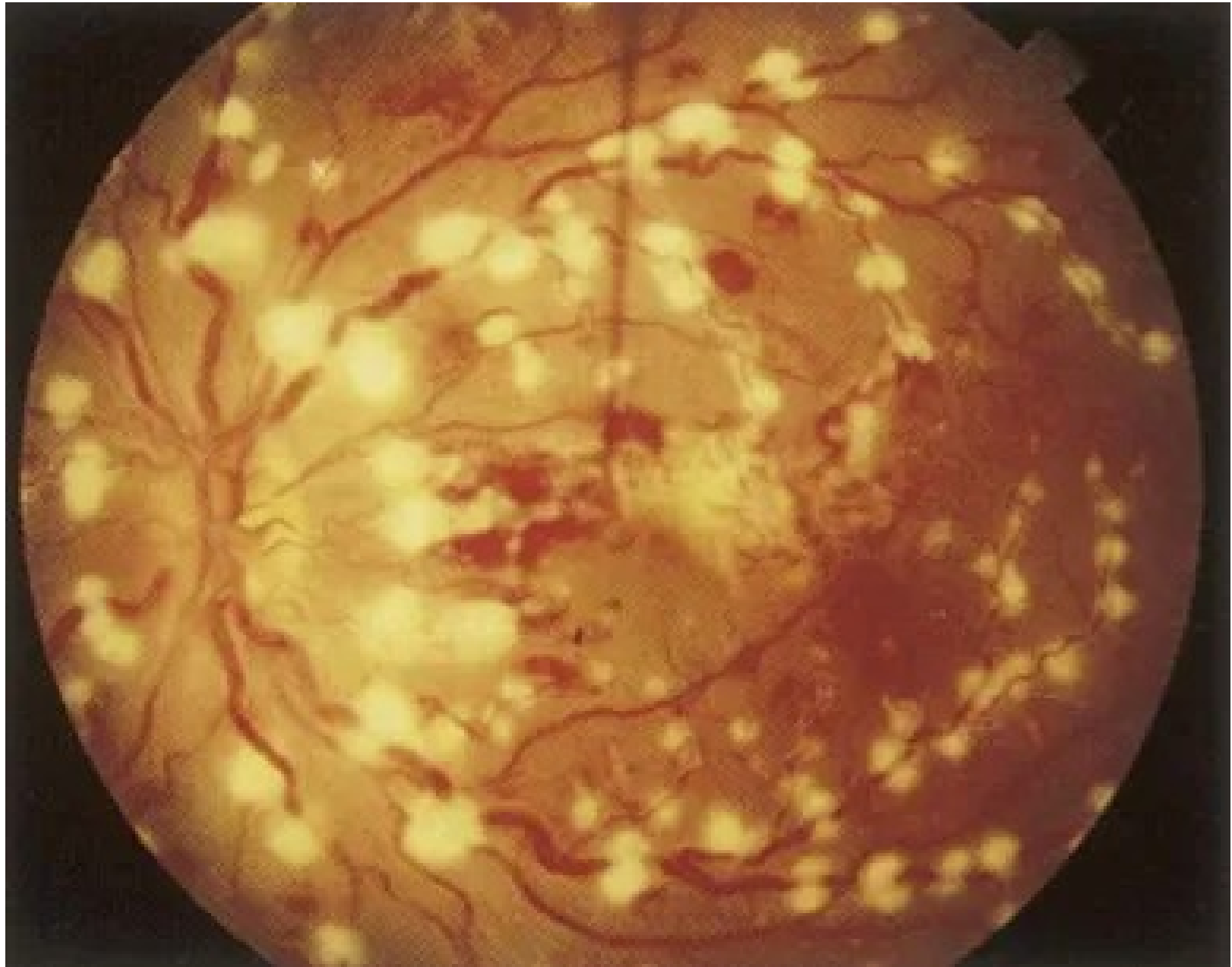


- retinal edema ( whitish appearance of the retina usually in acute vascular occlusion )

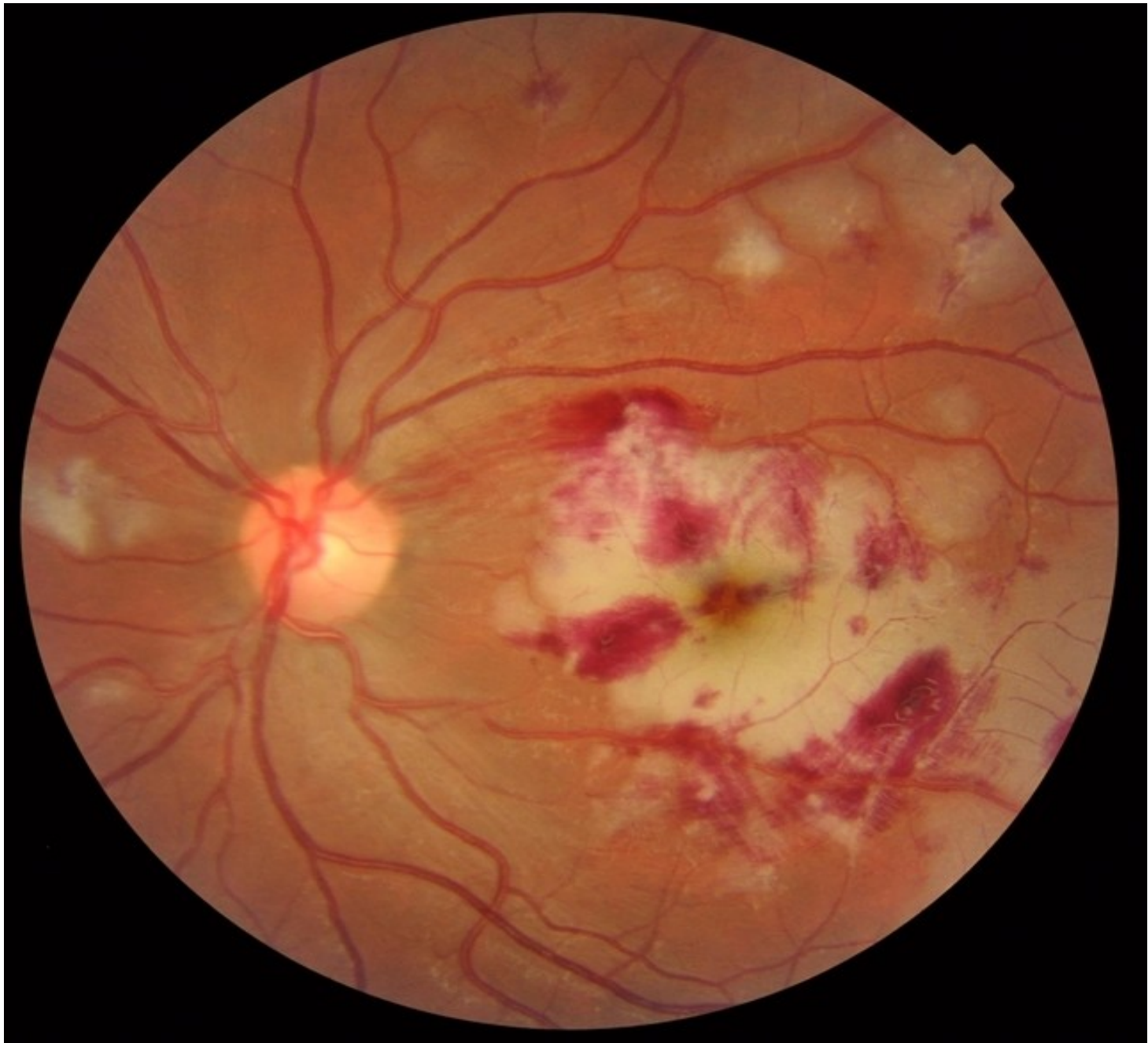


- retinal hemorrhages and cotton-wool spots ( frequently accompany retinal vasculitis, presumably related to the retinal ischemia produced by the inflammation)









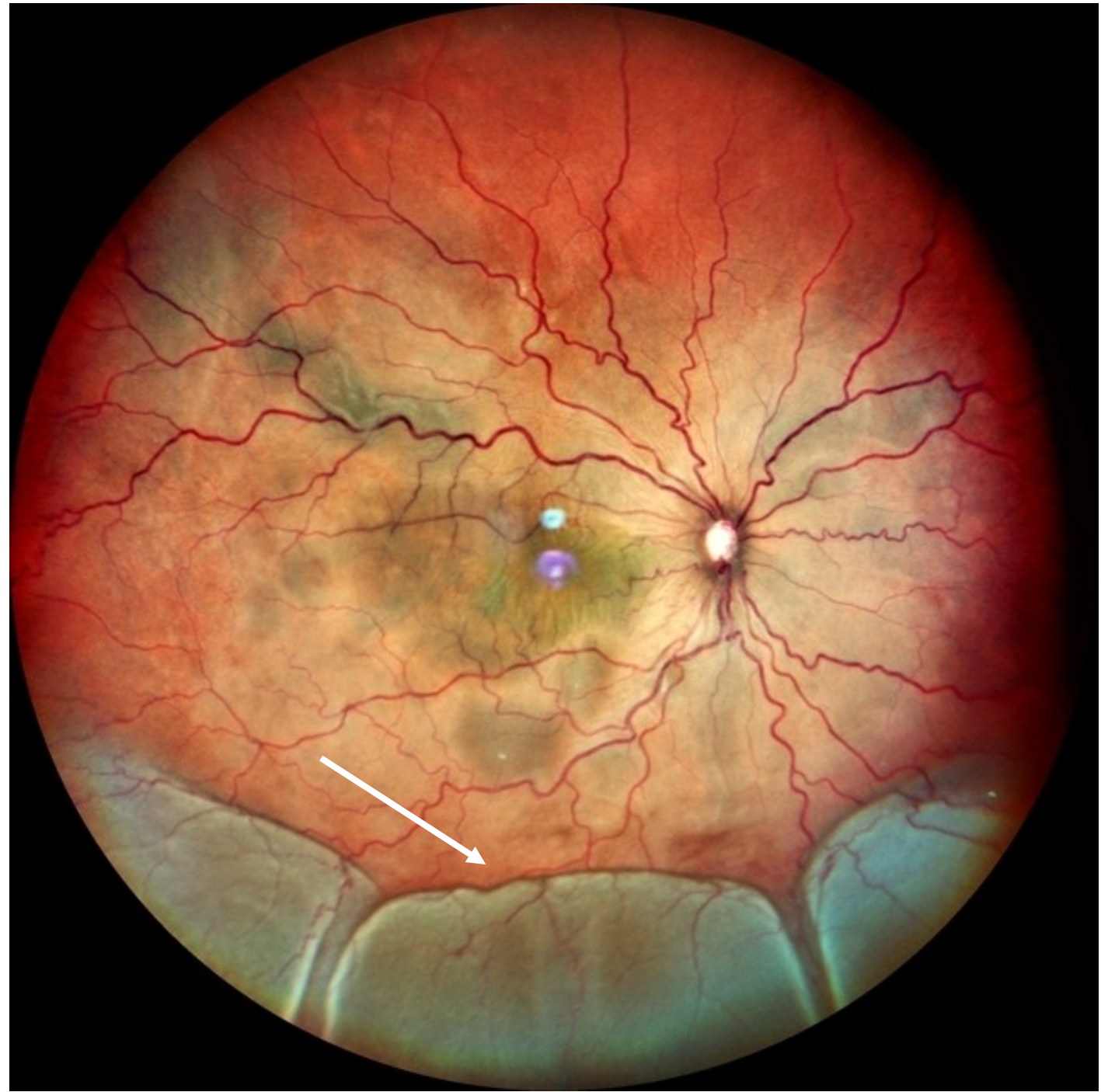


- retinal neovascularization  
( in occlusive retinal  
vascular disease or result  
of inflammatory stimuli in  
some diseases ;  
VKH, Behcet etc)



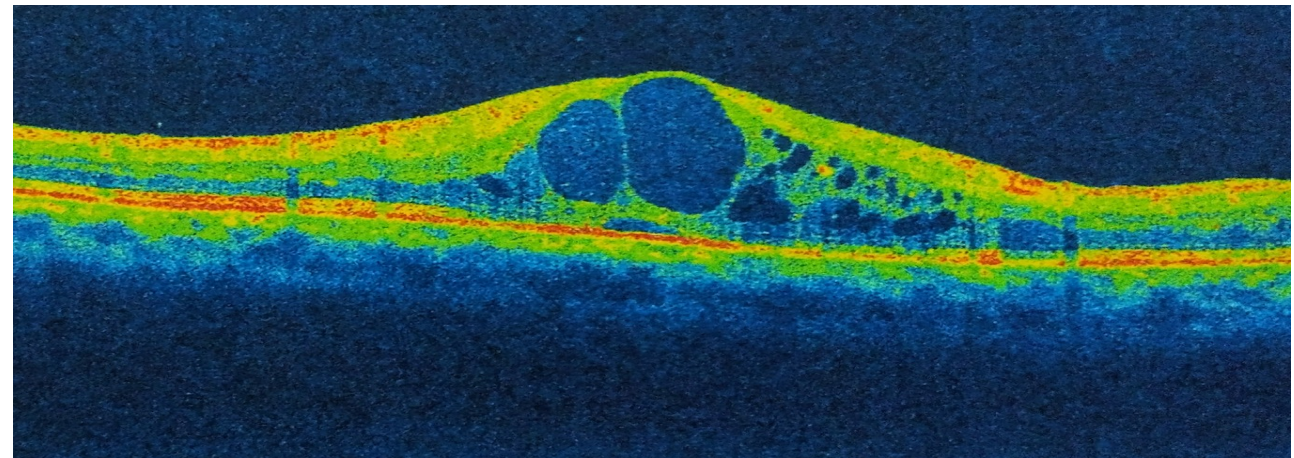
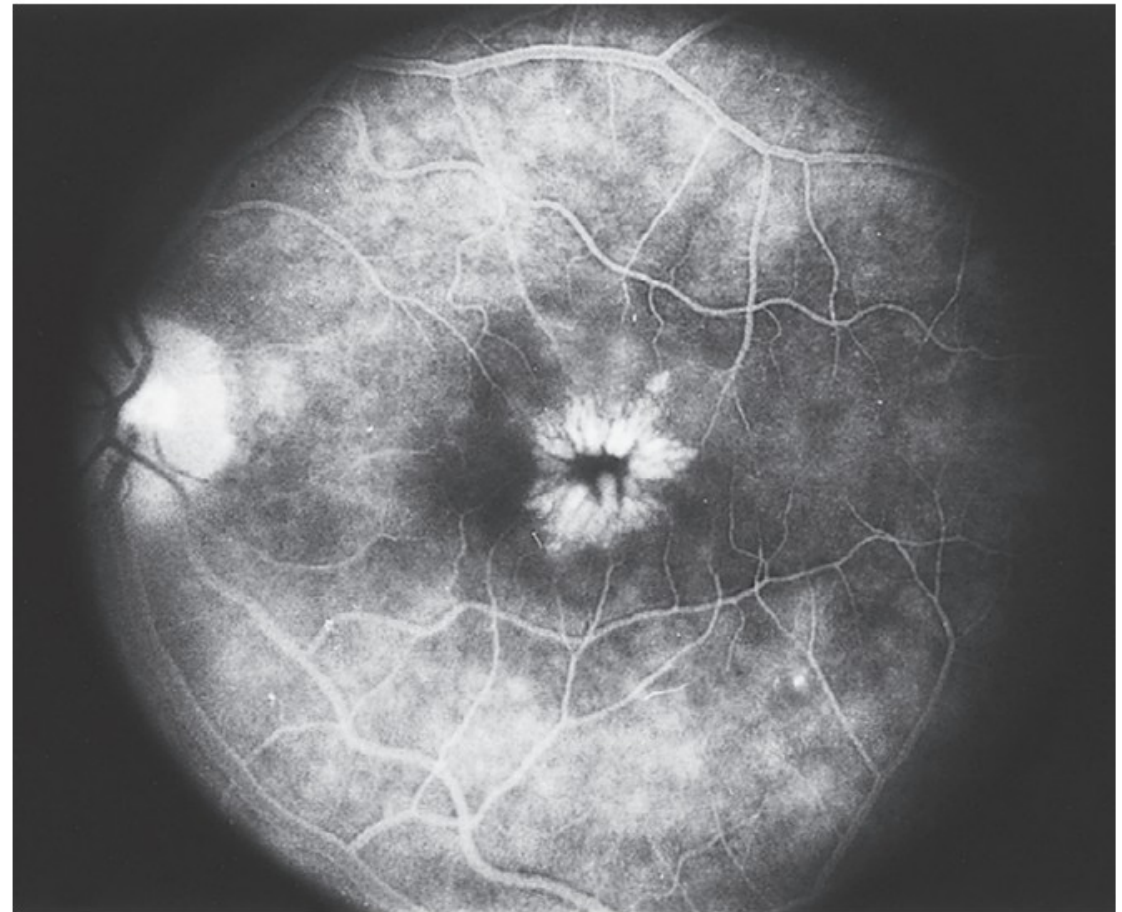
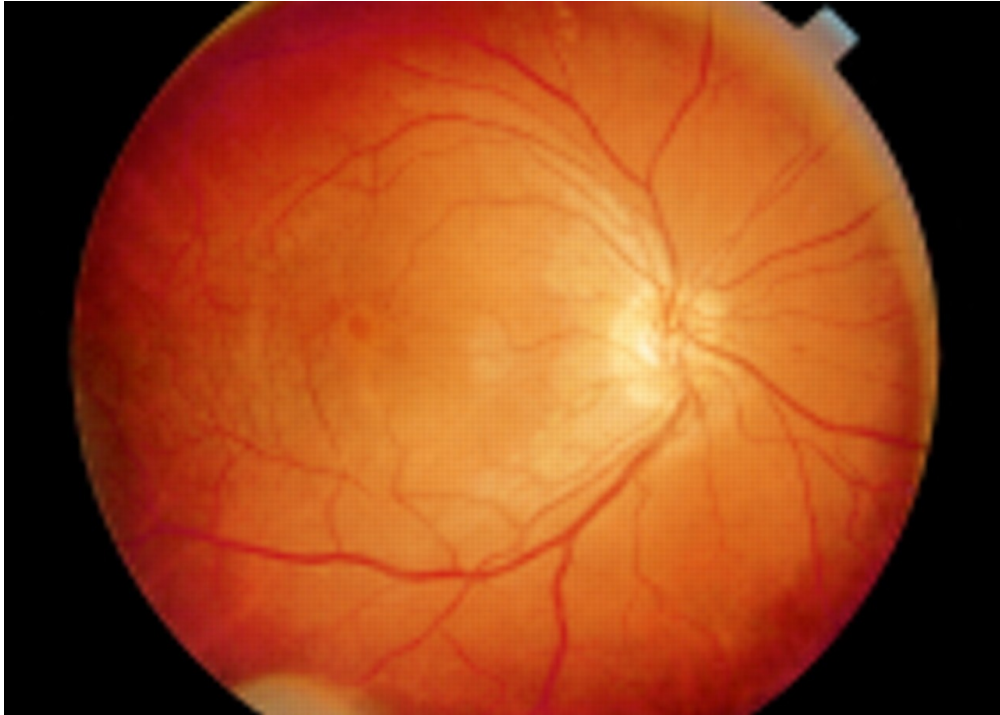


retinal detachment ( exudative  
– VKH disease )



- macular oedema

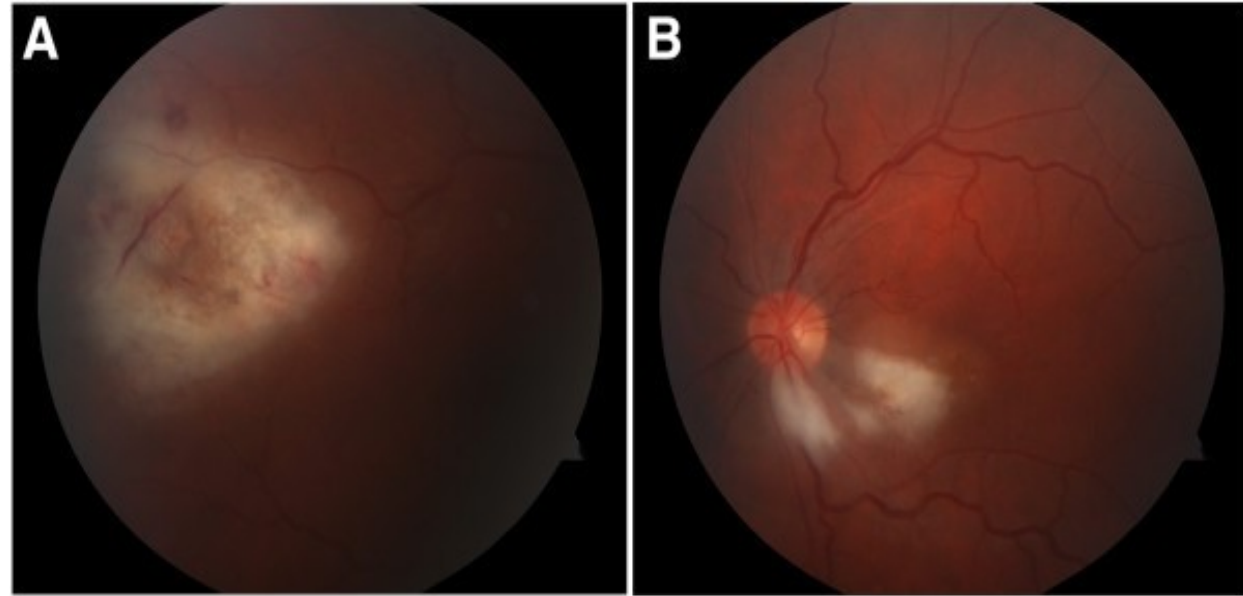
cystoid macular edema is a common retinal finding in patients with uveitis



*Fluorescein angiography can document the presence of macular edema more objectively and optical coherence tomography (OCT) is more frequently used to document the extent of macular edema*

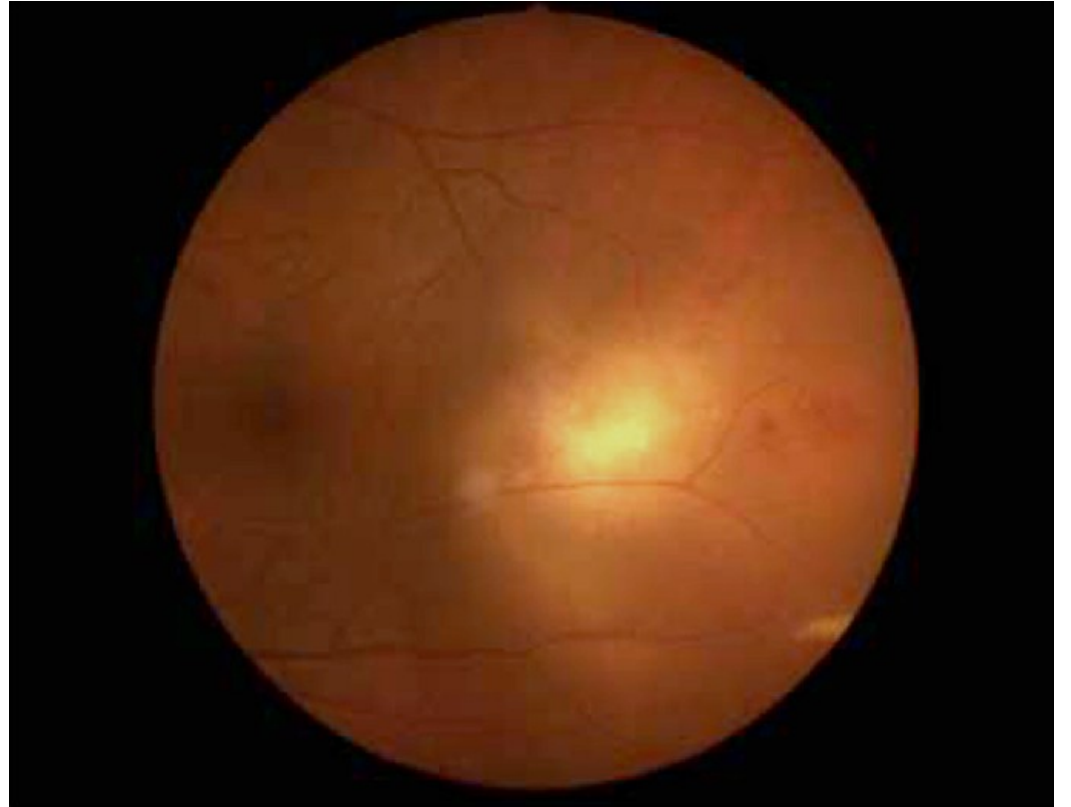
- retinitis

- cellular retinal infiltrates can be observed as **white areas** frequently have fuzzy edges, overlying vitreal cells, and surrounding retinal oedema



## Noninfective Causes of Retinitis

- Behcet disease (occlusive vasculitis)
- Sarcoidosis
- Other :
  - Systemic lupus erythematosus (SLE)
  - Churg–Strauss syndrome
  - Granulomatosis with polyangiitis





## Chorioretinal infiltrates

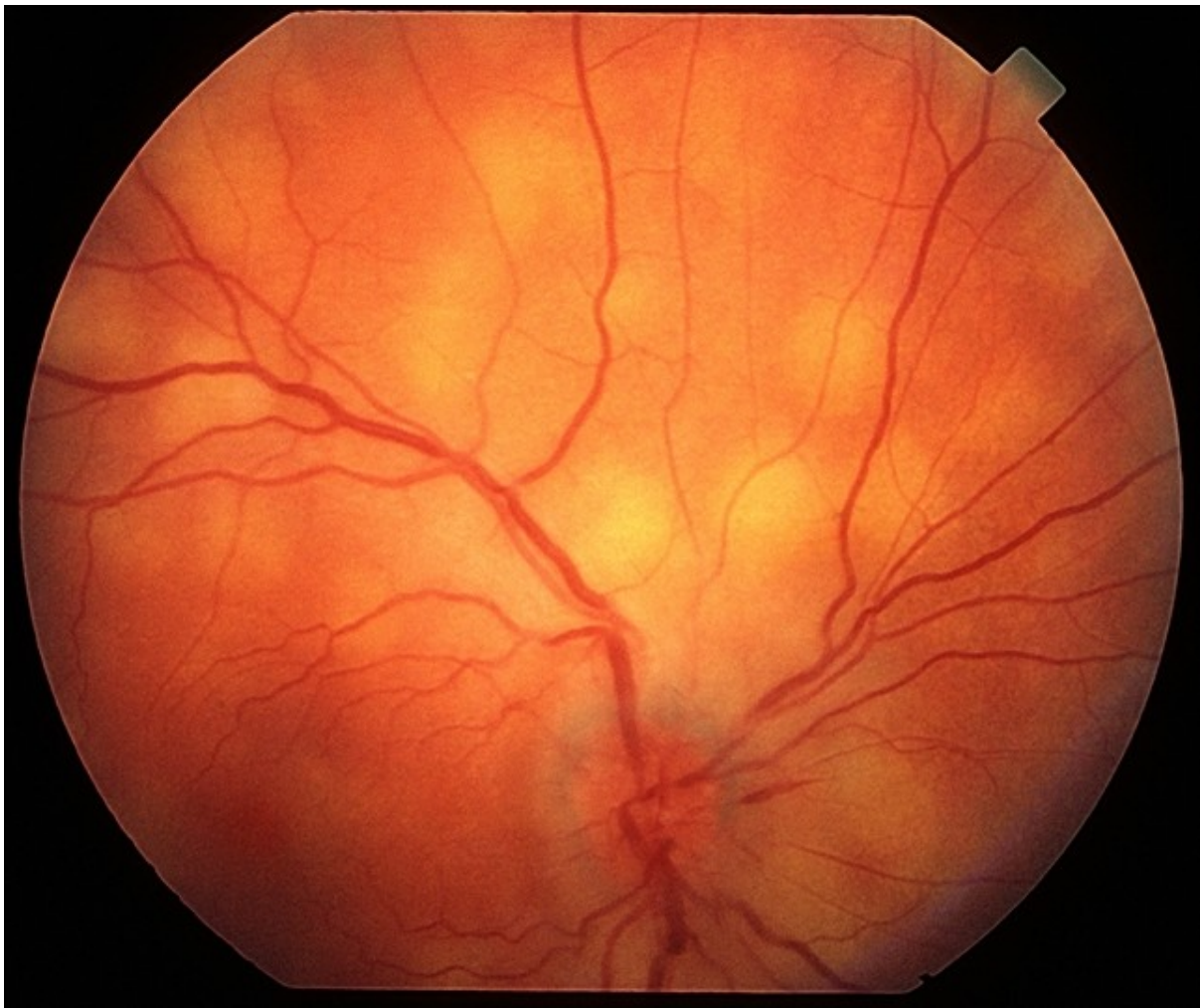
Cellular infiltrates within the choroid/outer retina

Single or multifocal

In many forms of posterior uveitis

Associated vitritis

Associated retinal oedema



# Causes of Intermediate Uveitis

Sarcoidosis

Inflammatory bowel disease

Multiple sclerosis

Lyme disease

Pars planitis<sup>a</sup>

## Causes of Posterior Uveitis

### Focal Retinitis

Toxoplasmosis  
Onchocerciasis  
Cysticercosis  
Masquerade syndromes

### Multifocal Retinitis

Syphilis  
Herpes simplex virus  
Cytomegalovirus  
Sarcoidosis  
Masquerade syndromes  
Candidiasis  
*Meningococcus*

### Focal Choroiditis

Toxocariasis  
Tuberculosis  
Nocardiosis  
Masquerade syndromes

### Multifocal Choroiditis

Histoplasmosis  
Sympathetic ophthalmia  
Vogt-Koyanagi-Harada syndrome  
Sarcoidosis  
Serpiginous choroidopathy  
Birdshot choroidopathy  
Masquerade syndromes (metastatic tumor)

## Causes of Panuveitis

Syphilis

Sarcoidosis

Vogt-Koyanagi-Harada syndrome

Infectious endophthalmitis

Behçet disease

## Causes and Associations of Retinal Vasculitis

Non infectious associations	Infectious agents
Behcet's disease	Mycobacterium Tuberculosis
Sarcoidosis	Treponema pallidum
Systemic lupus erythematosus	Toxoplasma gondii
Multiple sclerosis	Bartonella henselae
Seronegative arthropathies	Borrelia burgdorferi
Inflammatory bowel disease	Brucella
Sjogren's syndrome	Leptospira
Polyarteritis nodosa	Leptospira
Wegener granulomatosis	HIV
Relapsing polychondritis	HTLV1
Lymphoproliferative disorders	Herpesviridae – HSV, VZV, CMV, EBV
Drug induced	

*With funduscopy we can have a direct view of vessels....*

## Commoner causes of Periphlebitis and Arteritis

Venulitis ( periphlebitis )	Arteritis	Arteritis and Periphlebitis
Intermediate uveitis	Susac's syndrome	Systemic vasculitides *
Sarcoidosis	Systemic vasculitides * (ANCA-positive)	Toxoplasma chorioretinitis
Multiple sclerosis	Herpetic retinopathies	Syphilis
Inflammatory bowel disease	Toxoplasma chorioretinitis	Tuberculosis-Eale's disease
	Syphilis	
Seronegative arthropathies		

\* Systemic vasculitides include Churg-Strauss, Wegener's granulomatosis, polyarteritis nodosa and ANCA-positive vasculitides

# CASES

## Behcet's disease

Systemic vasculitis

Affects large & small vessels

Recurrent oral ulceration + at least two of :

- genital ulcers

- ocular involvement

- skin lesions

- a specific pathergy test which provokes pustular inflammation resulting from skin scratching

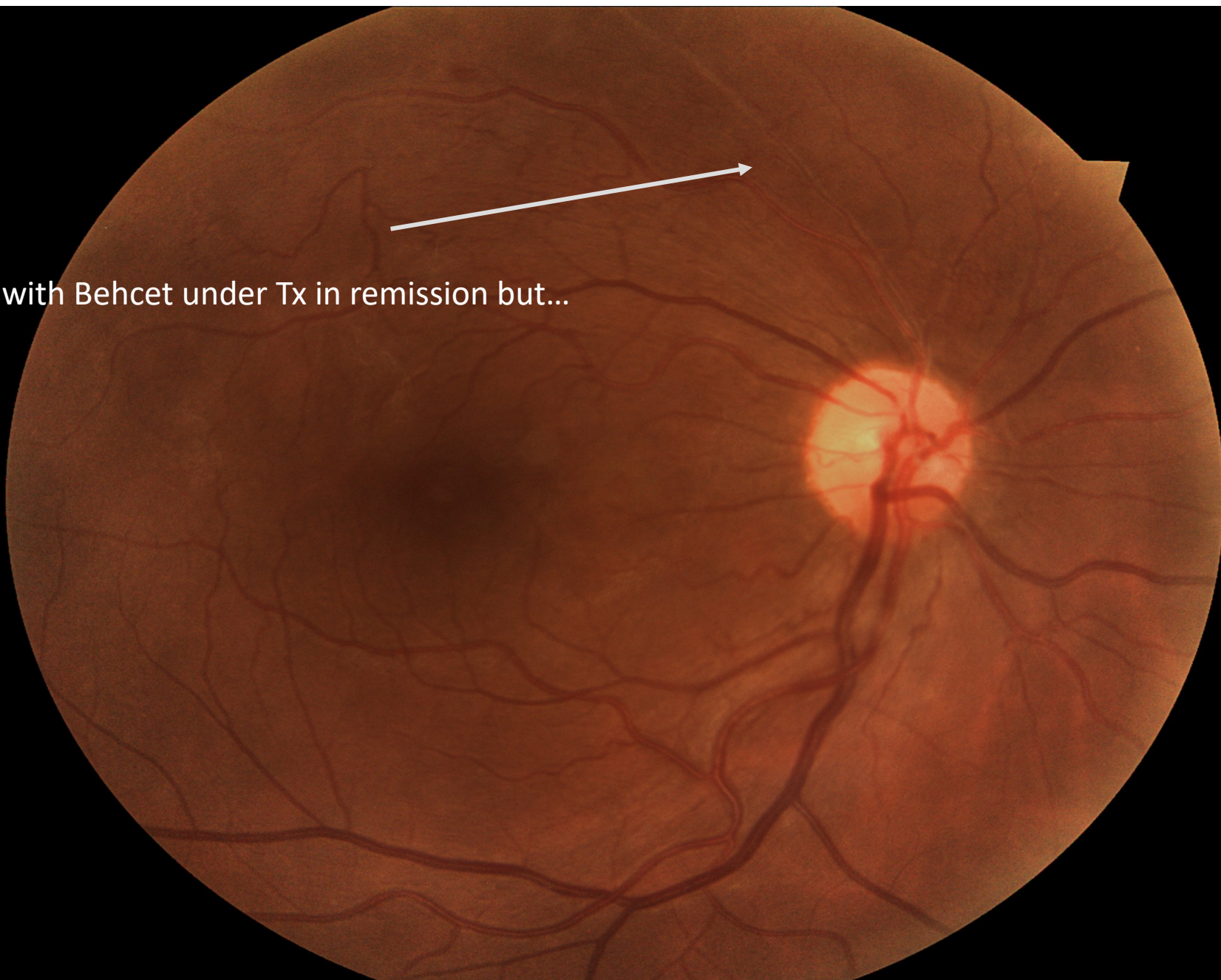


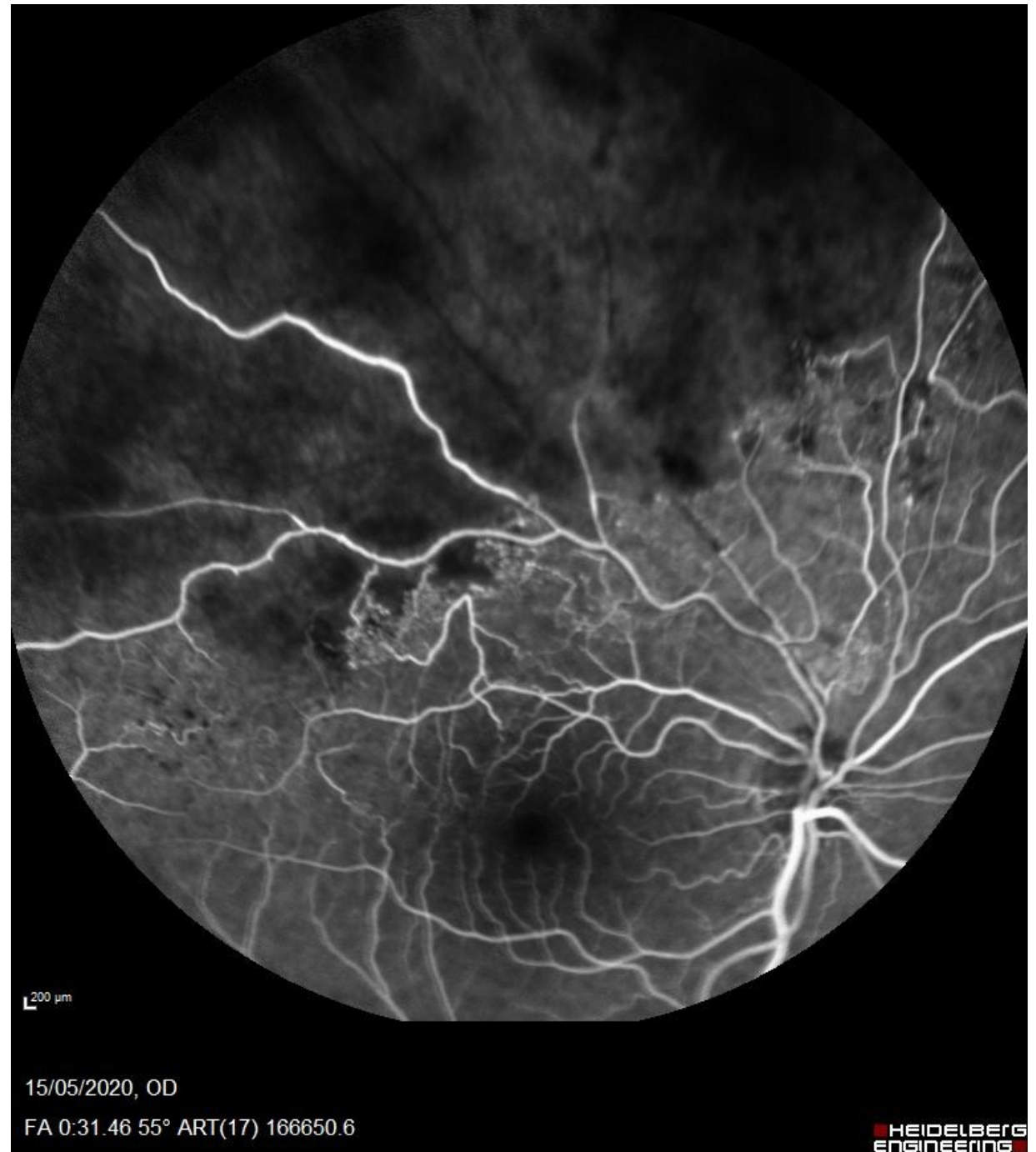
## Hallmark lesions of Behçet

- Vitritis
- Retinal vasculitis
- Retinitis
- Papillitis
- Panuveitis

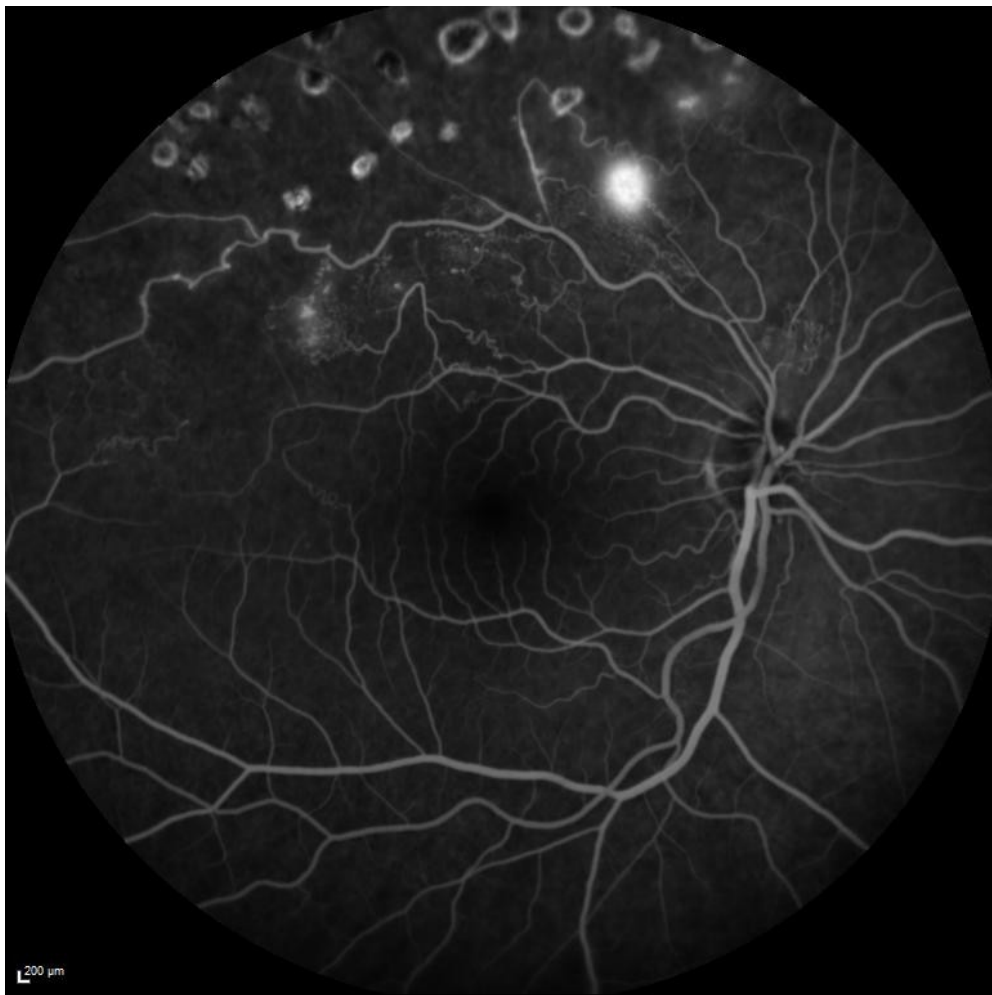


Patient with Behcet under Tx in remission but...





After escalation of Tx

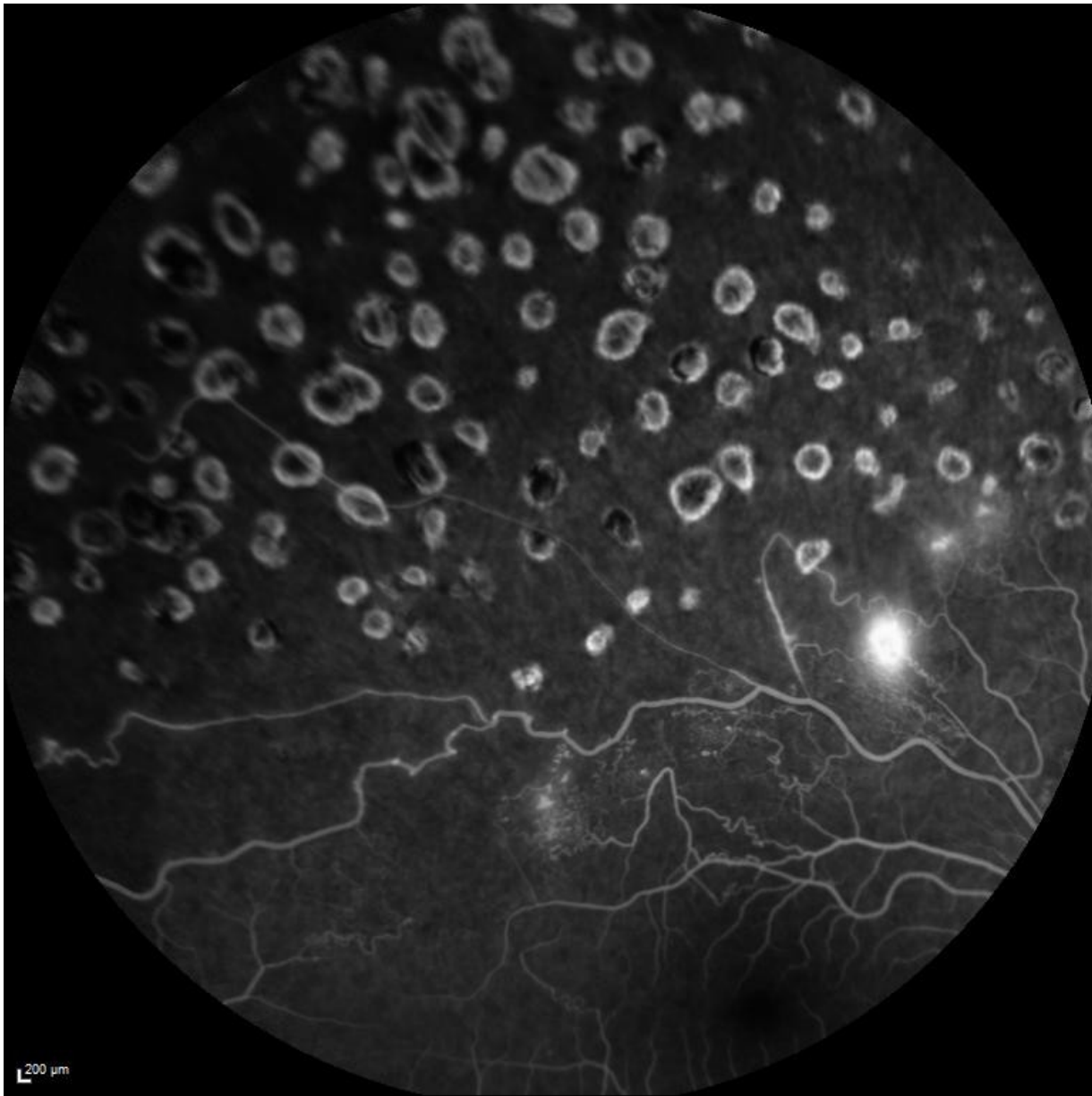


200  $\mu$ m

26/06/2023, OD

FA 1:07.38 55° ART(97) 262332.2

HEIDELBERG  
ENGINEERING



200  $\mu$ m

26/06/2023, OD

FA 1:22.11 55° ART(29) 262332.3

HEIDELBERG  
ENGINEERING

CMV retinitis in a  
immunosuppressed  
patient for  
autoimmune disease



RG OptomapPlus  
Oct 18, 2023 10:04 AM  
Image: 1  
4000  
4000

Laterality: R  
Red: 50%  
Green: 50%

After IV Tx ...

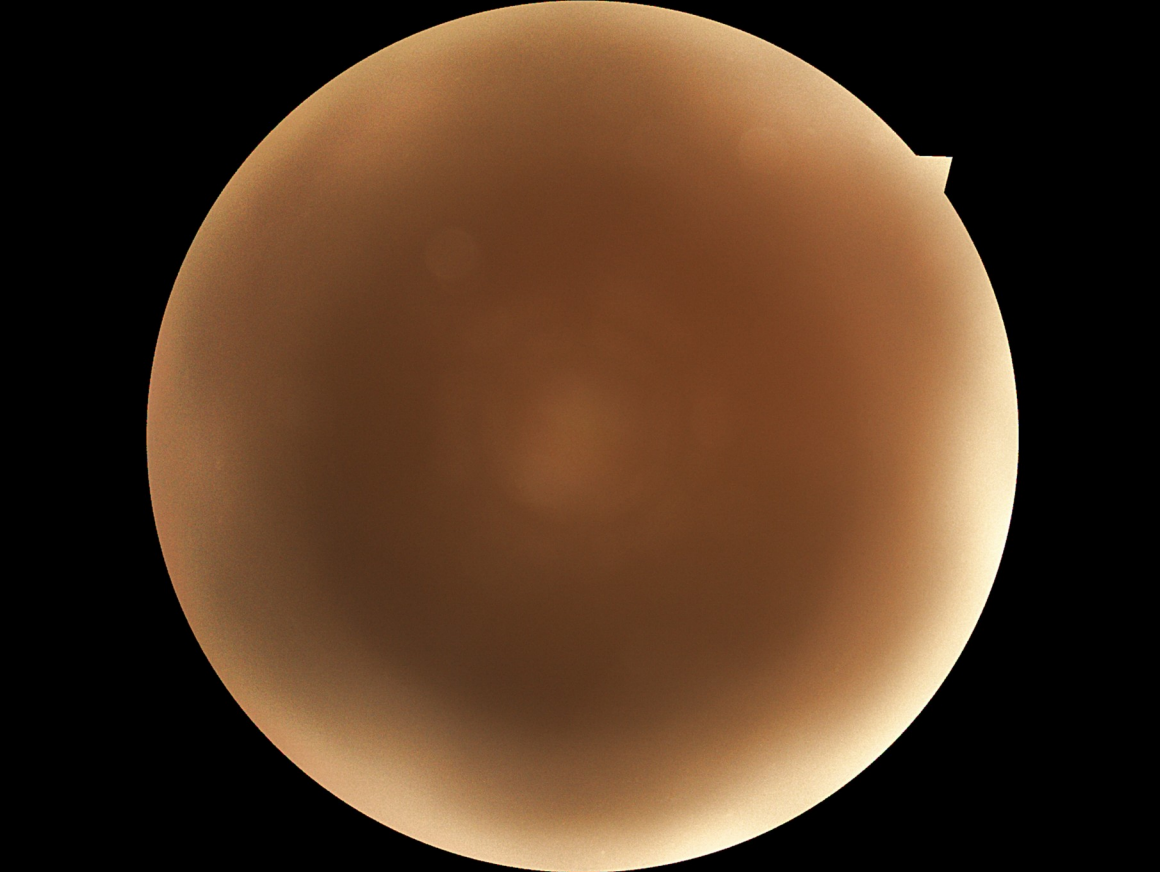
Zoom: 1.50  
Presentation: Multiple

1/3



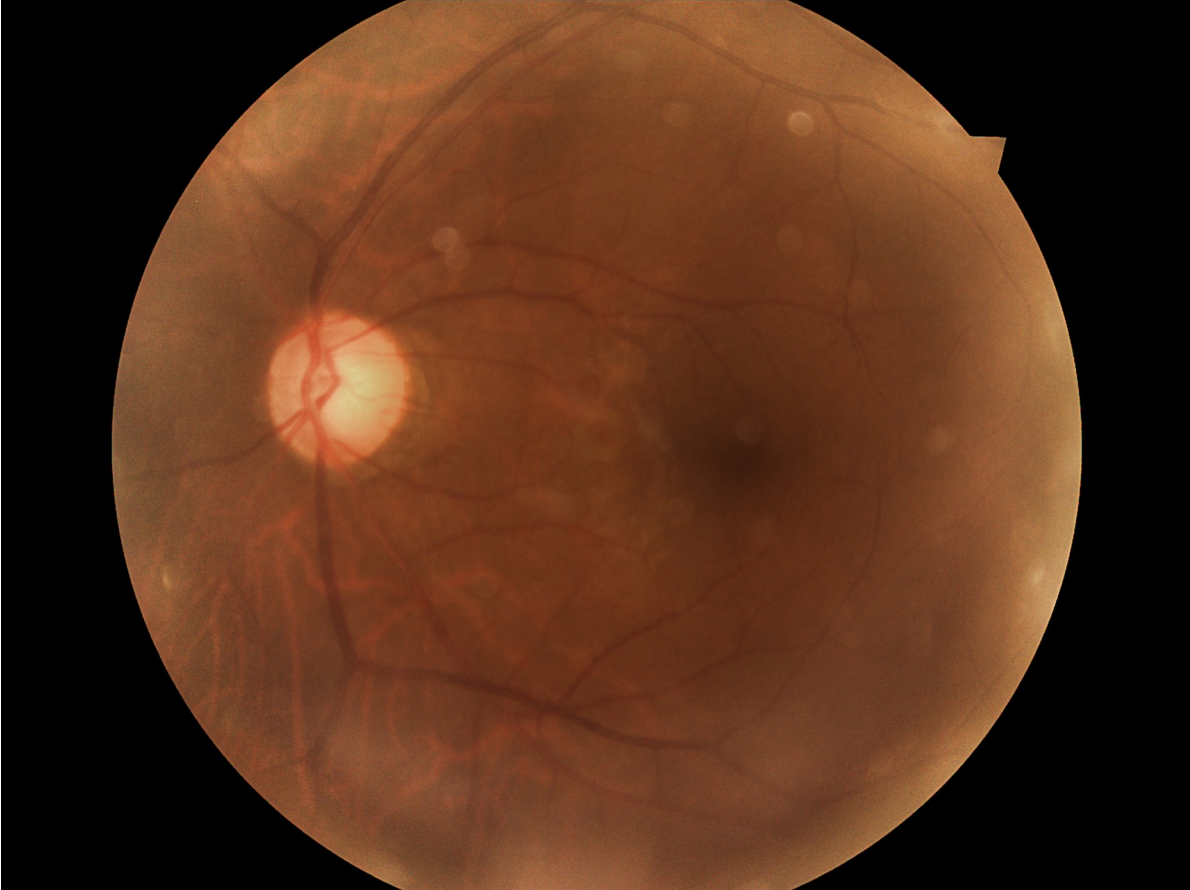
Choroidal  
granuloma in  
sarcoidosis





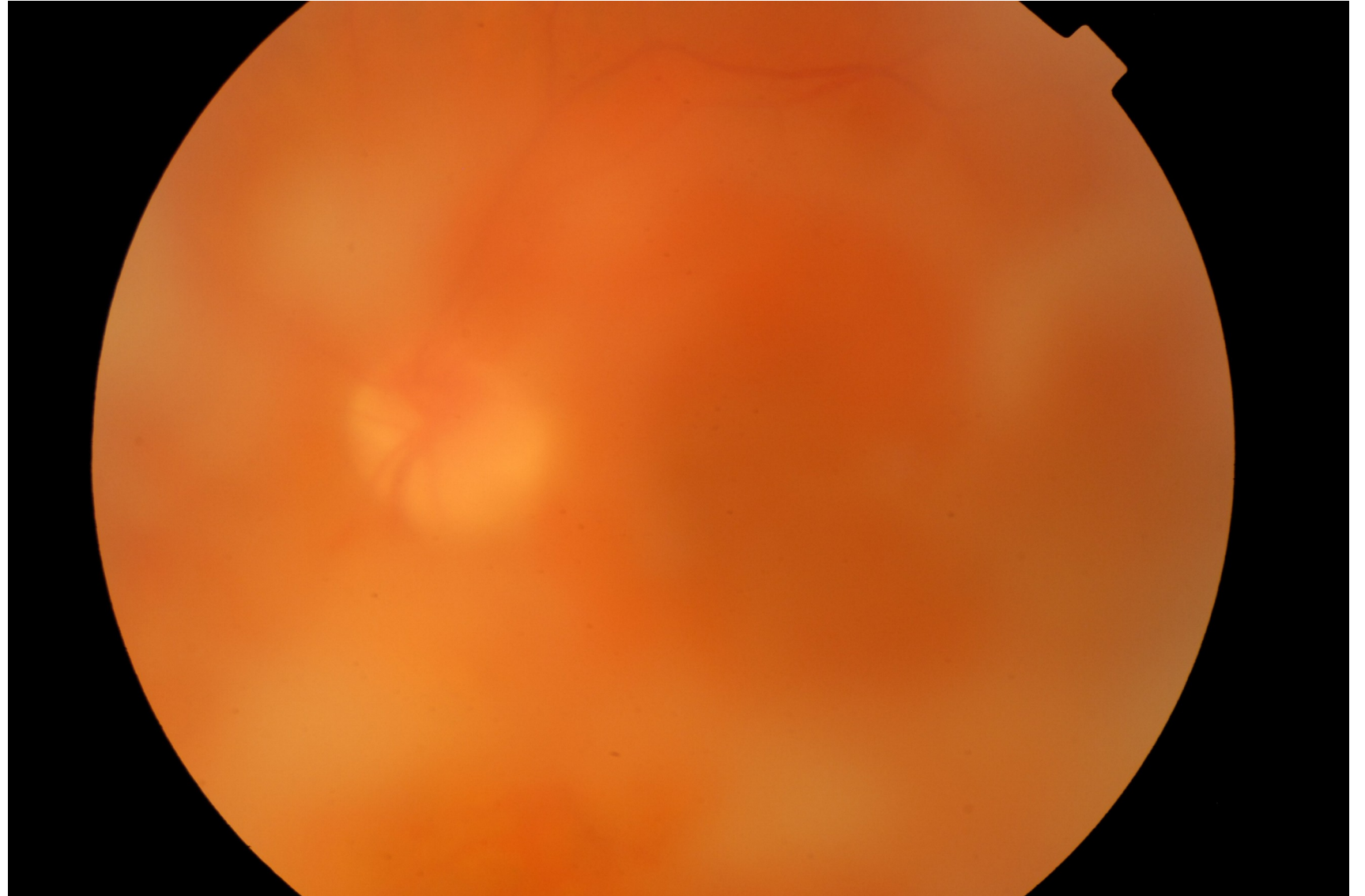
Dense vitreous haze in a patient with uveitis  
Bioscore 5 – no fundal view

Normal left eye



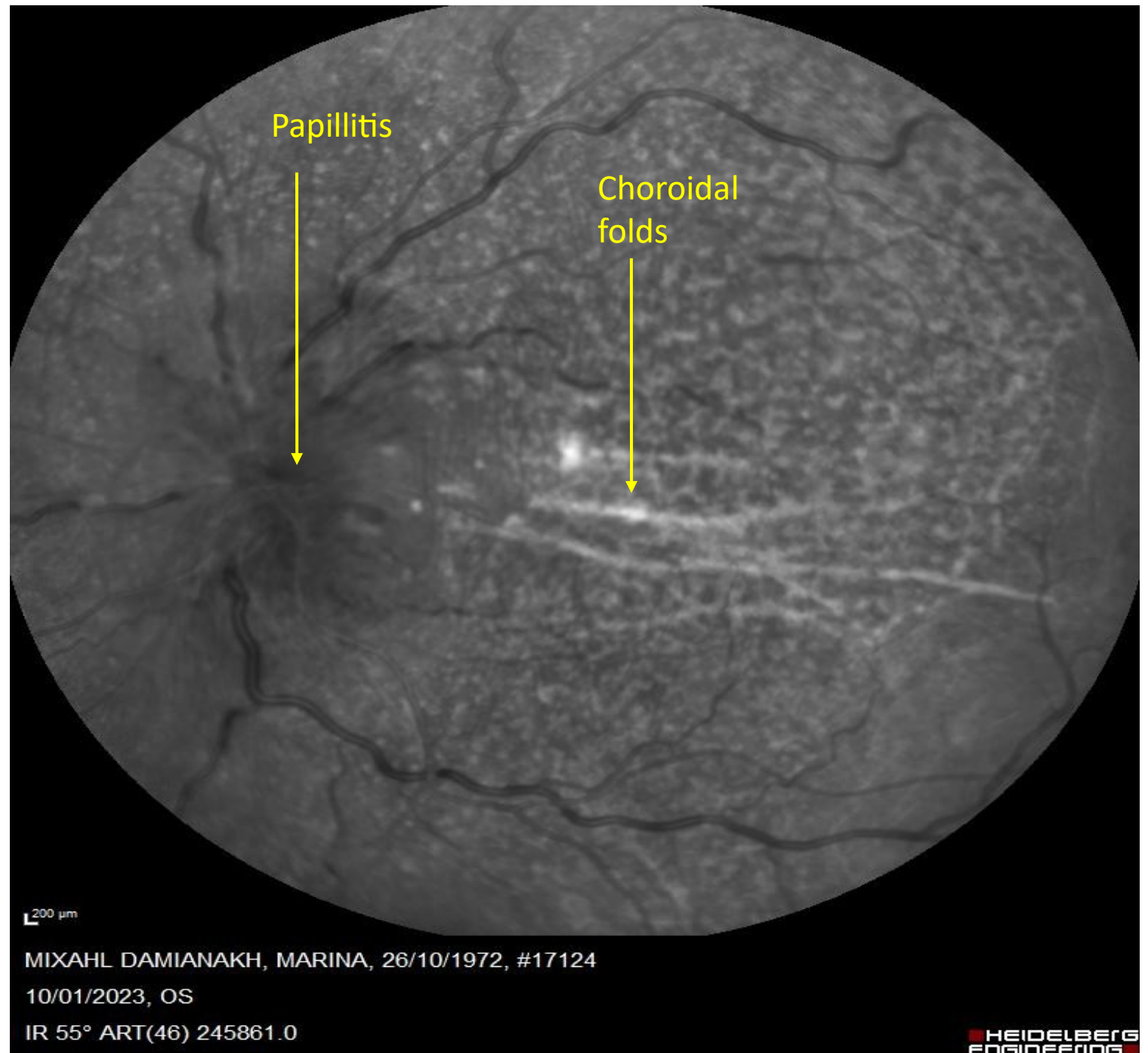


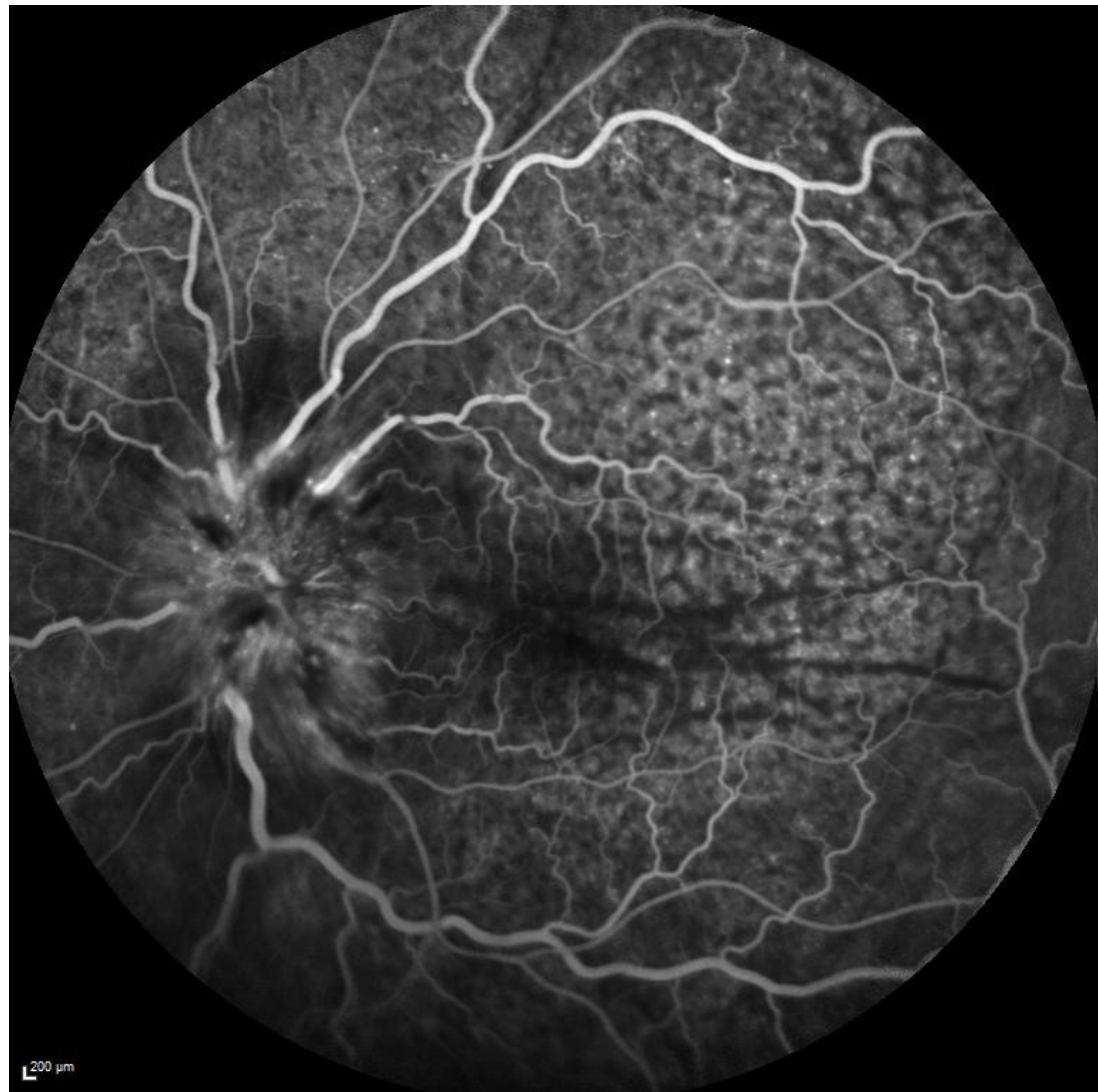
Severe vitritis /  
biosocre 4



Infrared photo of left retina in a patient with posterior scleritis associated with rheumatoid arthritis

Treated with immunosuppression by rheumatologists





200  $\mu$ m

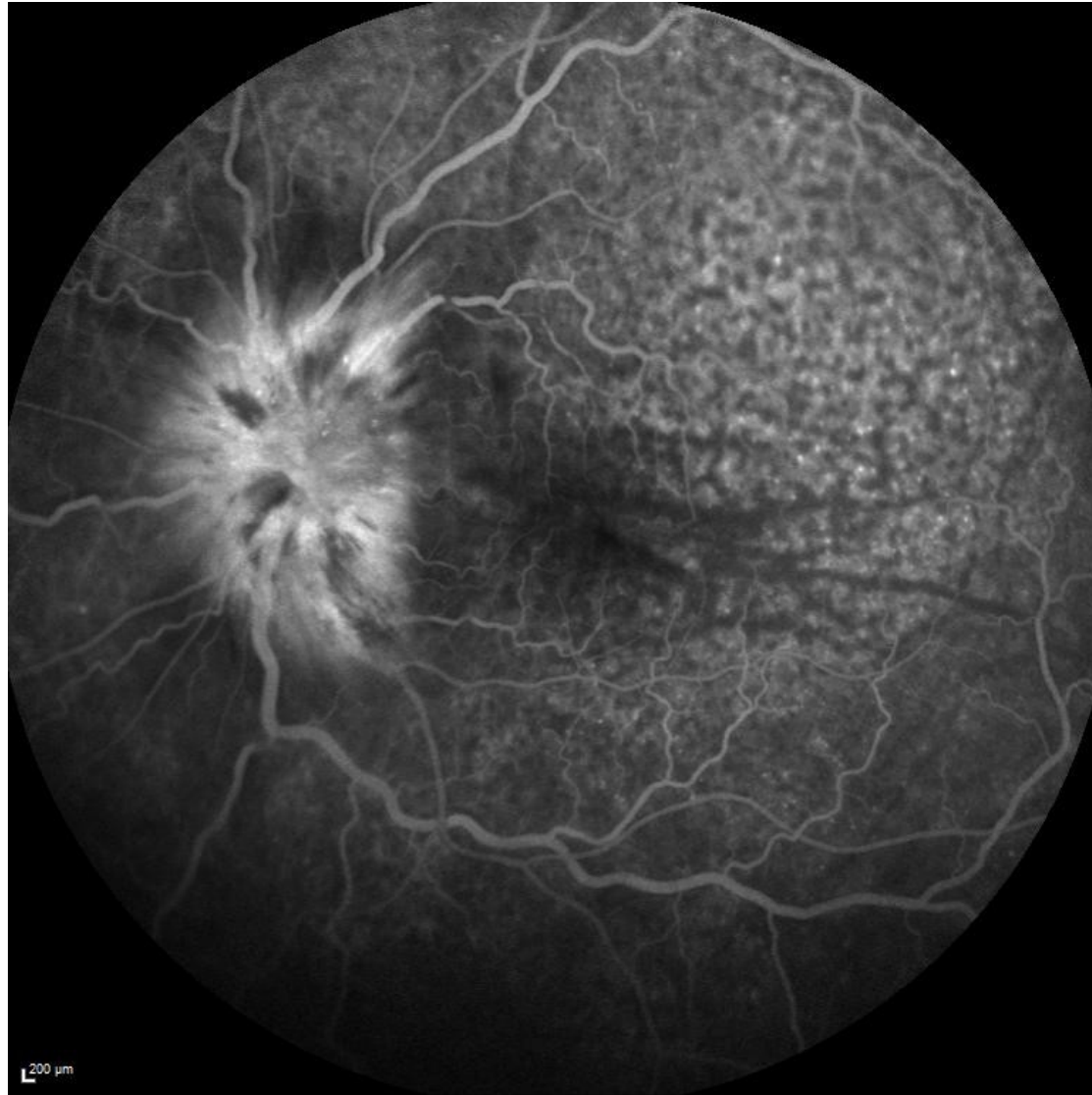


200  $\mu$ m

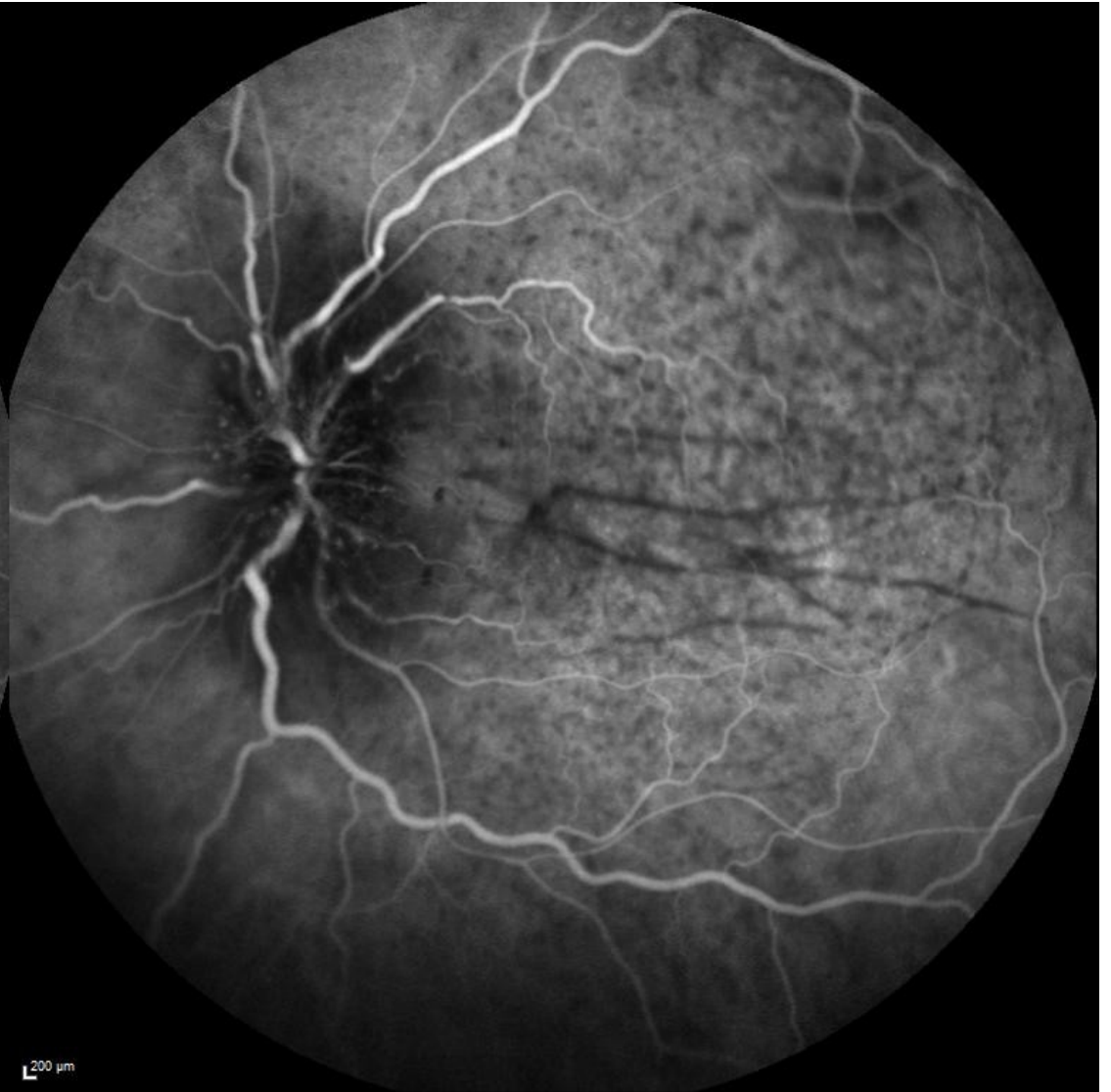
MIXAHL DAMIANAKH, MARINA, 26/10/1972, #17124

10/01/2023, OS

FA&ICGA 1:25.51 55° ART(22) 1:25.48 55° ART(22) 245864.0



200 µm



200 µm

MIXAHL DAMIANAKH, MARINA, 26/10/1972, #17124

10/01/2023, OS

FA&ICGA 5:48.57 55° ART(16) 5:48.53 55° ART(16) 245866.0

Pale optic disc in a patient with chronic uveitis



Papilledema in a young boy with autoimmune disorder





Optic disc oedema in  
patient with anterior  
uveitis associated  
with RA

Bilateral grade IV  
hypertensive retinopathy  
in an adolescent with

- ✓ Optic disc edema
- ✓ Macular edema
- ✓ Retinal arteriolar  
narrowing
- ✓ Elschnig spots
- ✓ Upper peripheral  
serous retinal  
detachment

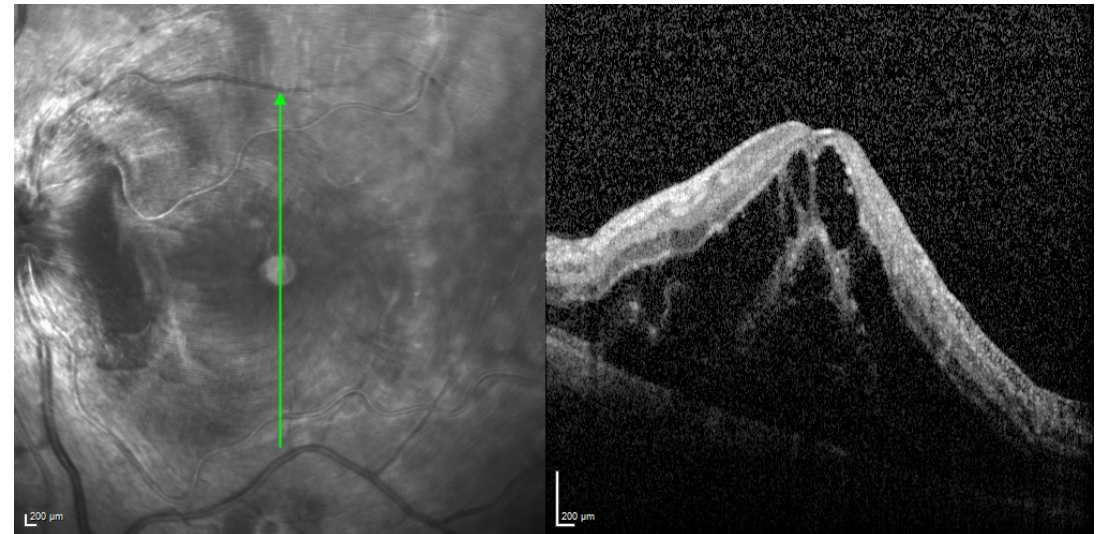
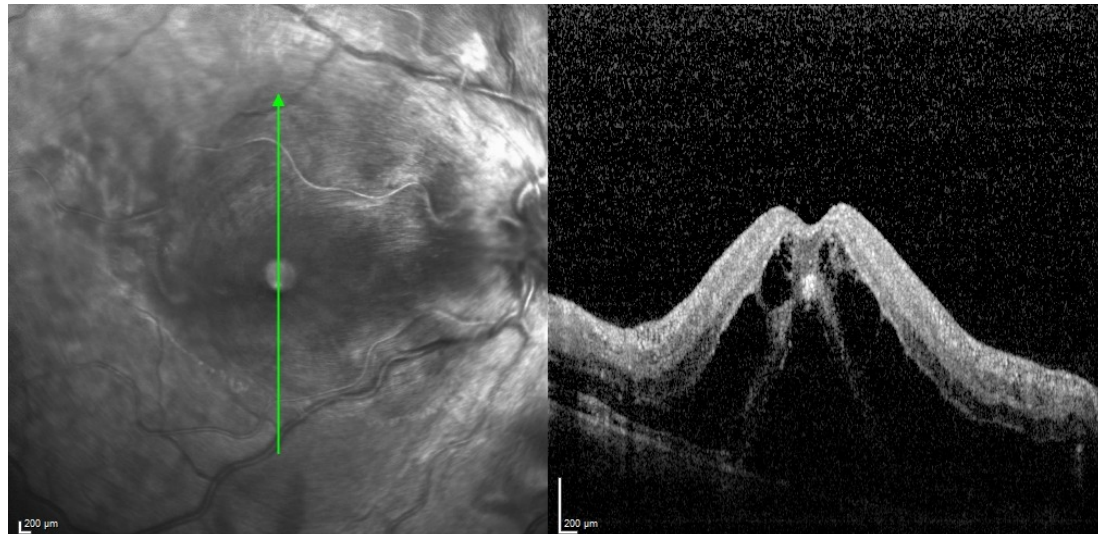
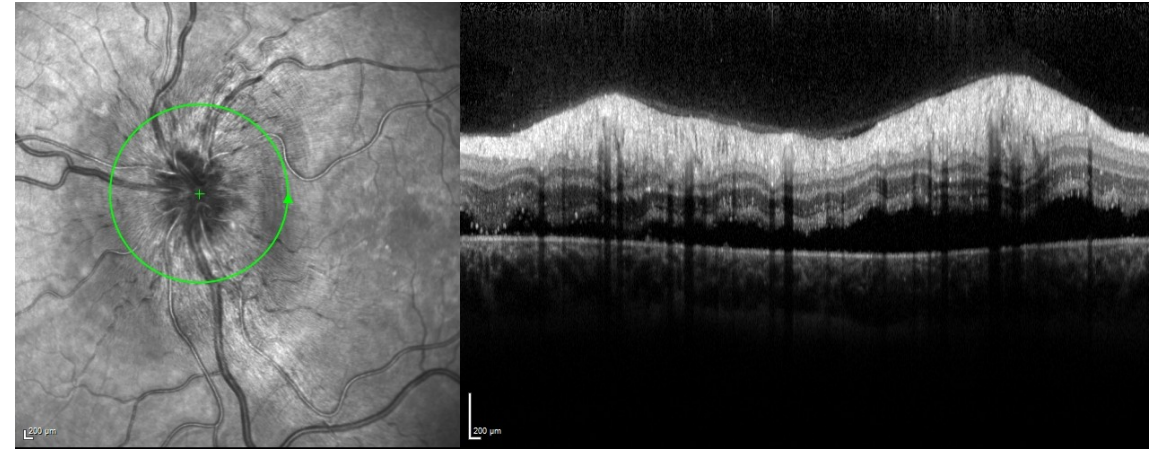
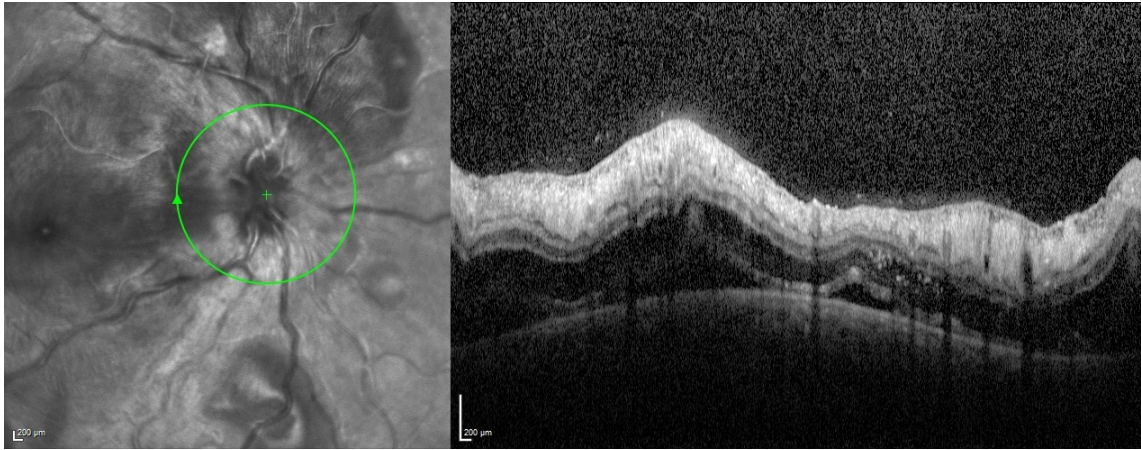




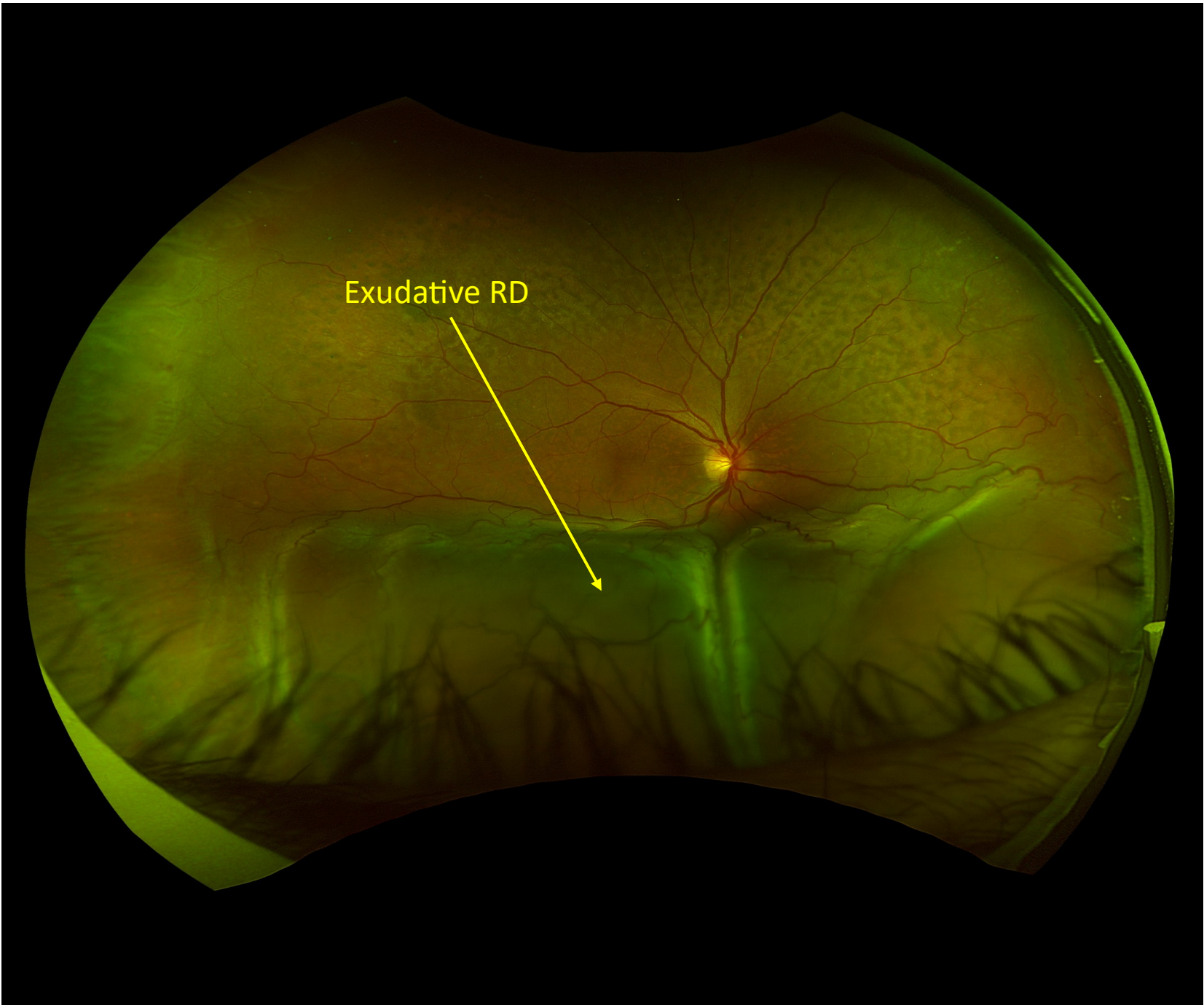
Blood pressure:  
220/170 mmHg

Complement 3  
Glomerulonephritis



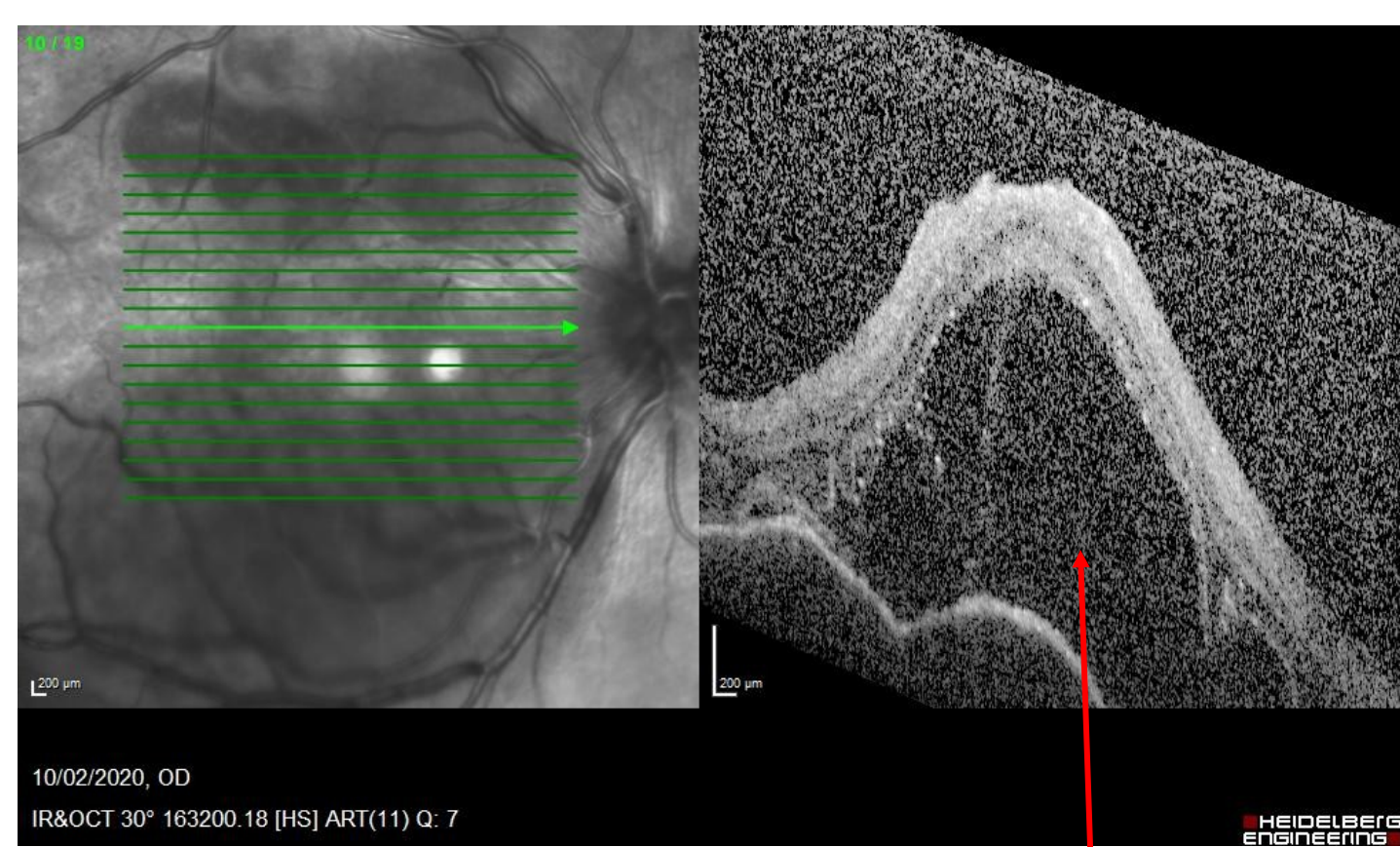


Exudative retinal detachment in a presumed Harada Disease



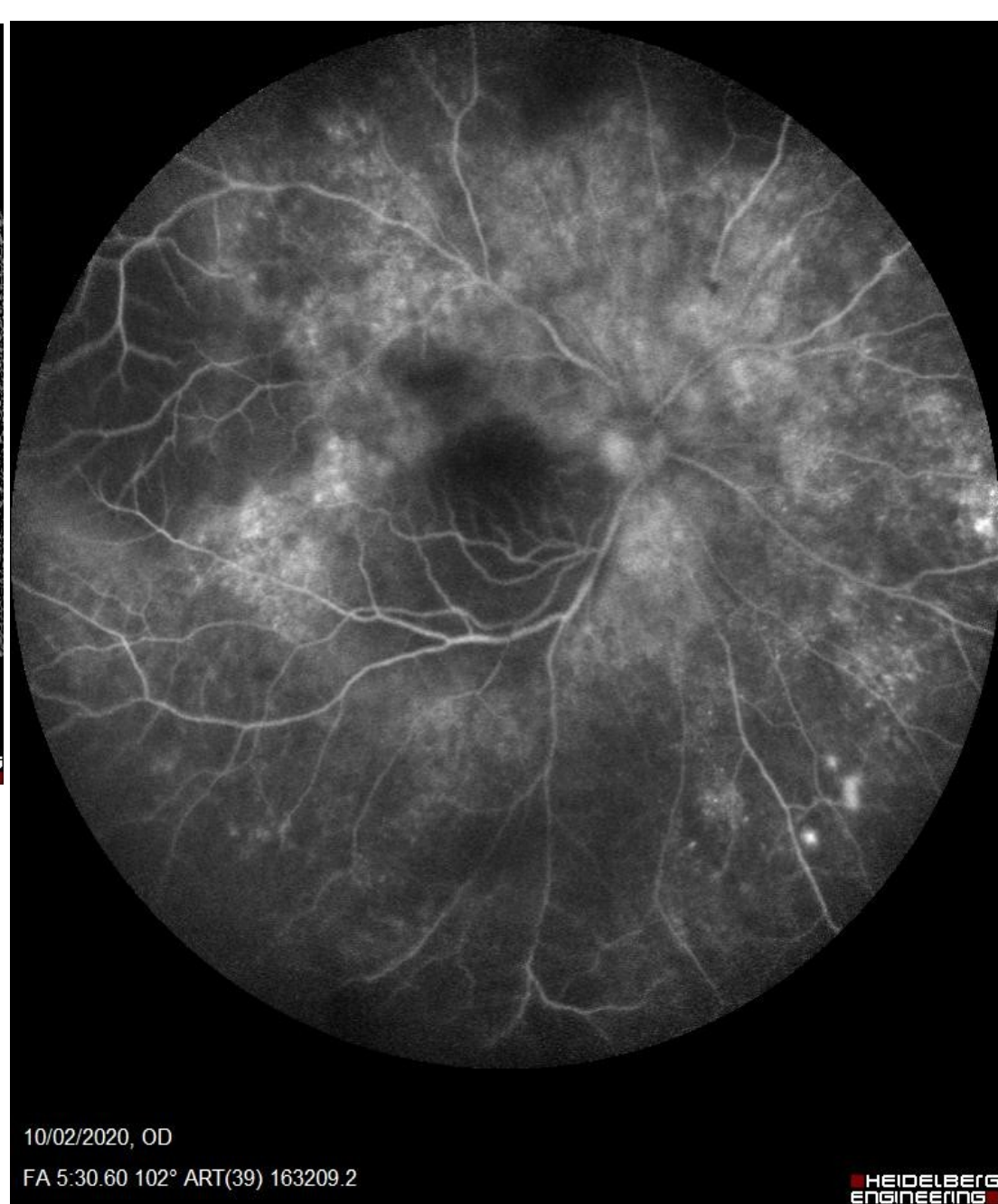
Previous Pt post Tx  
with oral steroids

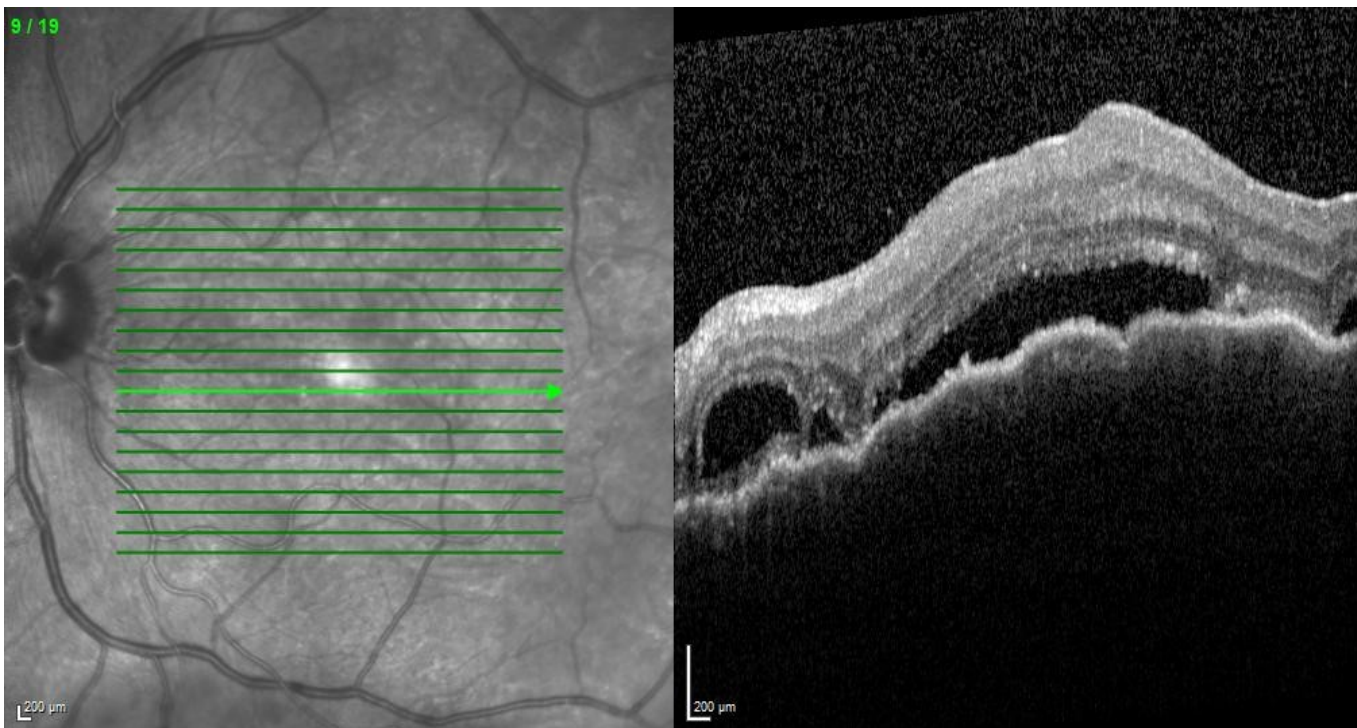




Subretinal fluid

VKH disease in a 70  
years old man

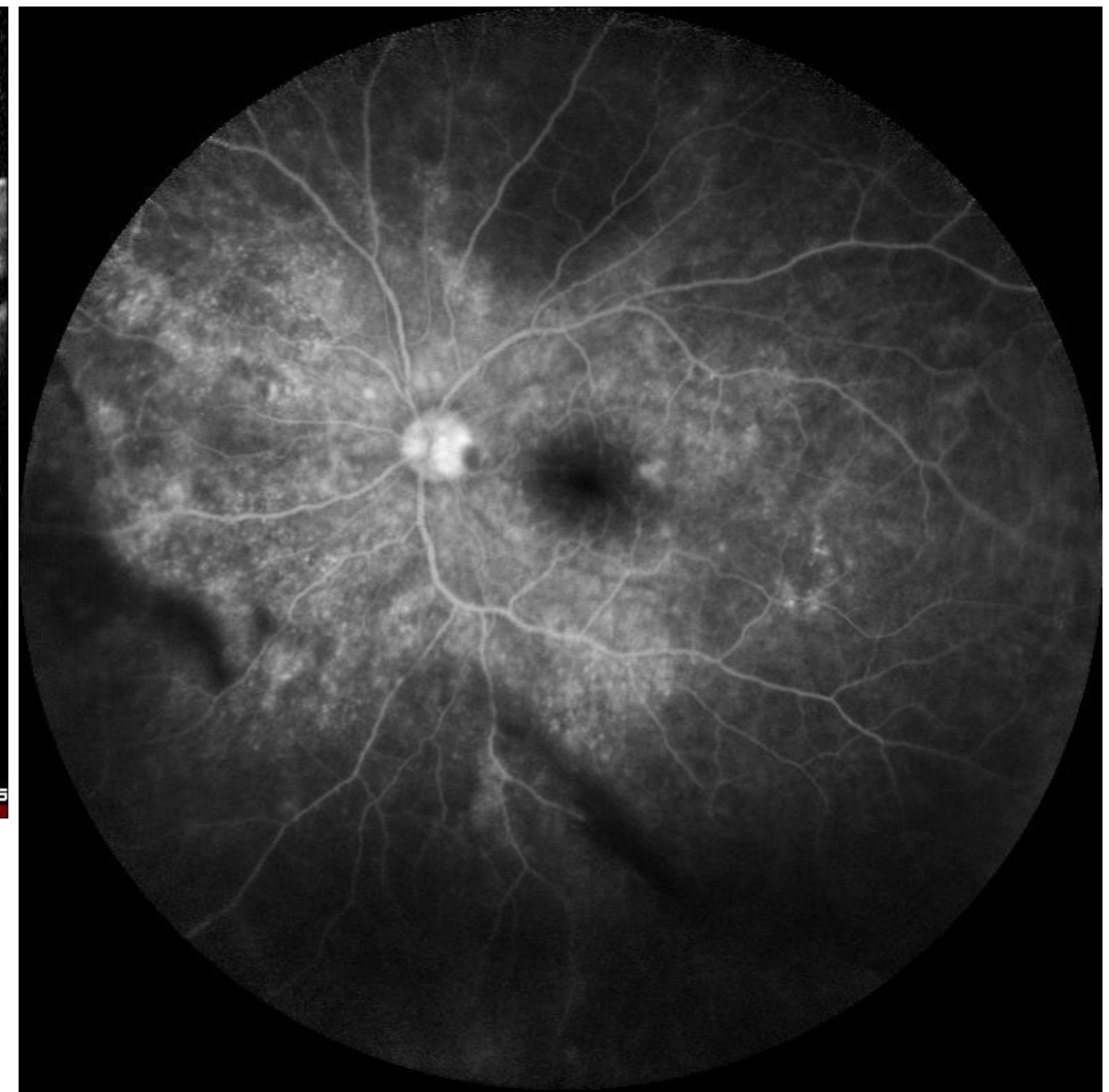




10/02/2020, OS  
IR&OCT 30° ART 163201.16 [HS] ART(11) Q: 22

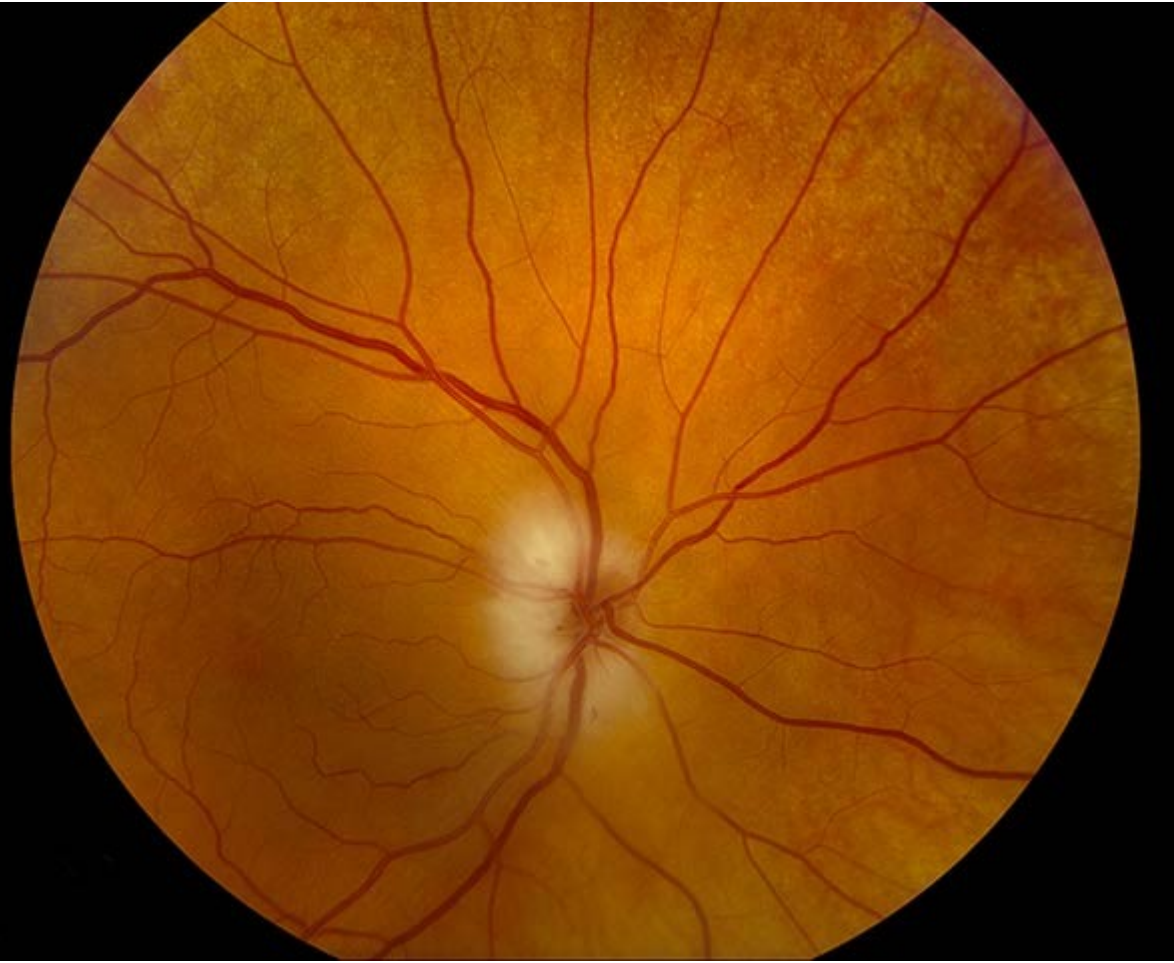
HEIDELBERG  
ENGINEERING

VKH in a 70 years old man



10/02/2020, OS  
FA 5:59.84 102° ART(45) 163210.0

HEIDELBERG  
ENGINEERING



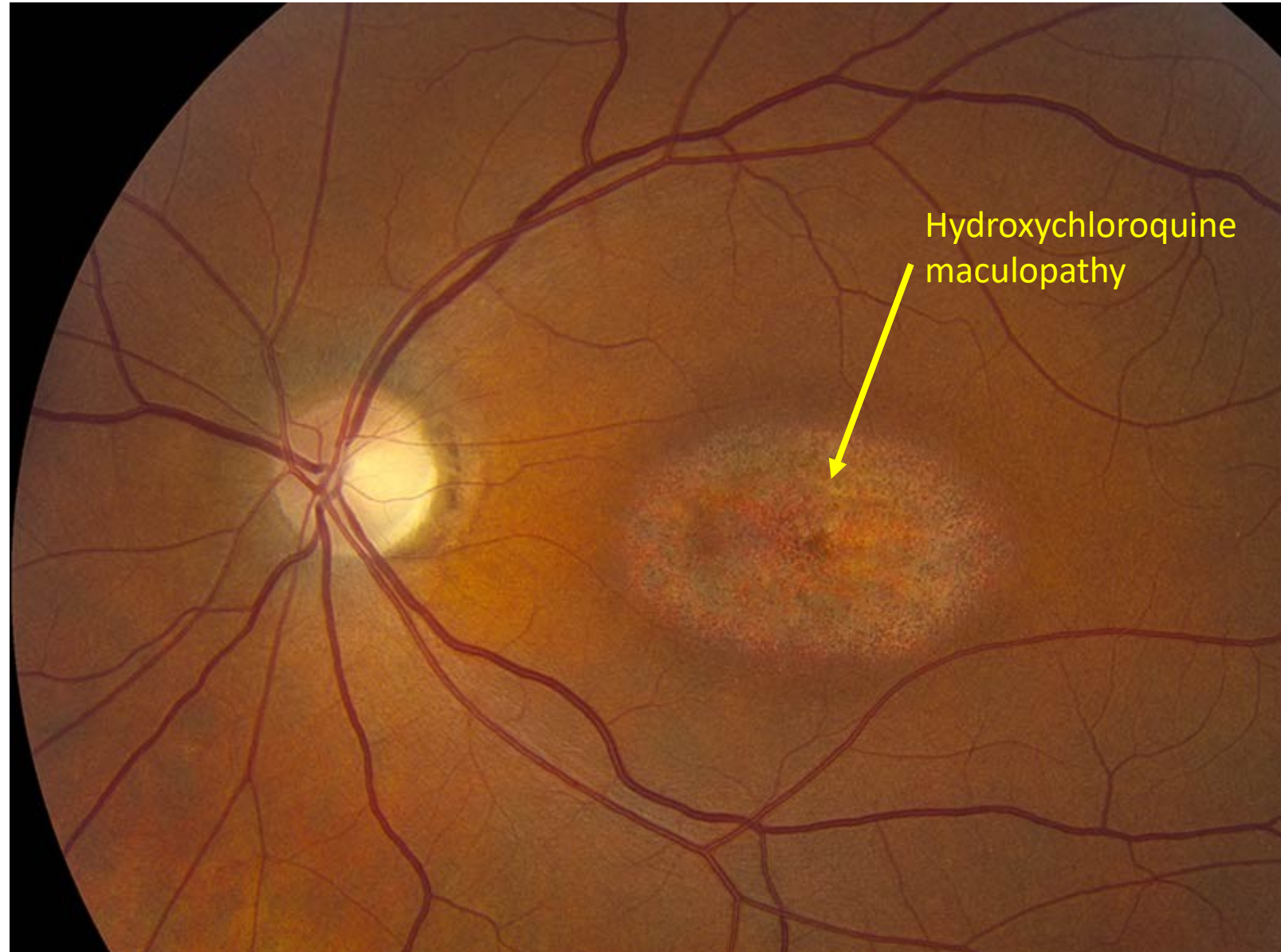
## Anterior Ischemic Optic Neuropathy (AAION) in GCA

- Profound sudden vision loss
- Jaw claudication
- Scalp tenderness
- Headache
- Elevated ESR, CRP

## Retinal toxicity of drugs used in rheumatology...



Regular monitoring  
is necessary as  
retinal damage is  
irreversible...





## FUNDUS EXAMINATION

- ✓ can provide important information and guide clinicians towards the right diagnosis
- ✓ is necessary in many cases for determining if an autoimmune disease with ocular involvement is under remission or not thus tailoring treatment
- ✓ ophthalmologists and rheumatologists frequently work together as most autoimmune diseases affect the eyes
- ✓ basic funduscopy can be done by a non ophthalmologist and in cases where a consultation by an eye specialist is not available, can provide help to the clinician for diagnostic dilemmas



*Low cost handheld fundus cameras can provide clinicians useful information before referral to eye specialists*





*Or more  
expensive non  
mydriatric  
cameras...*

Thank you....

