

CME in a child

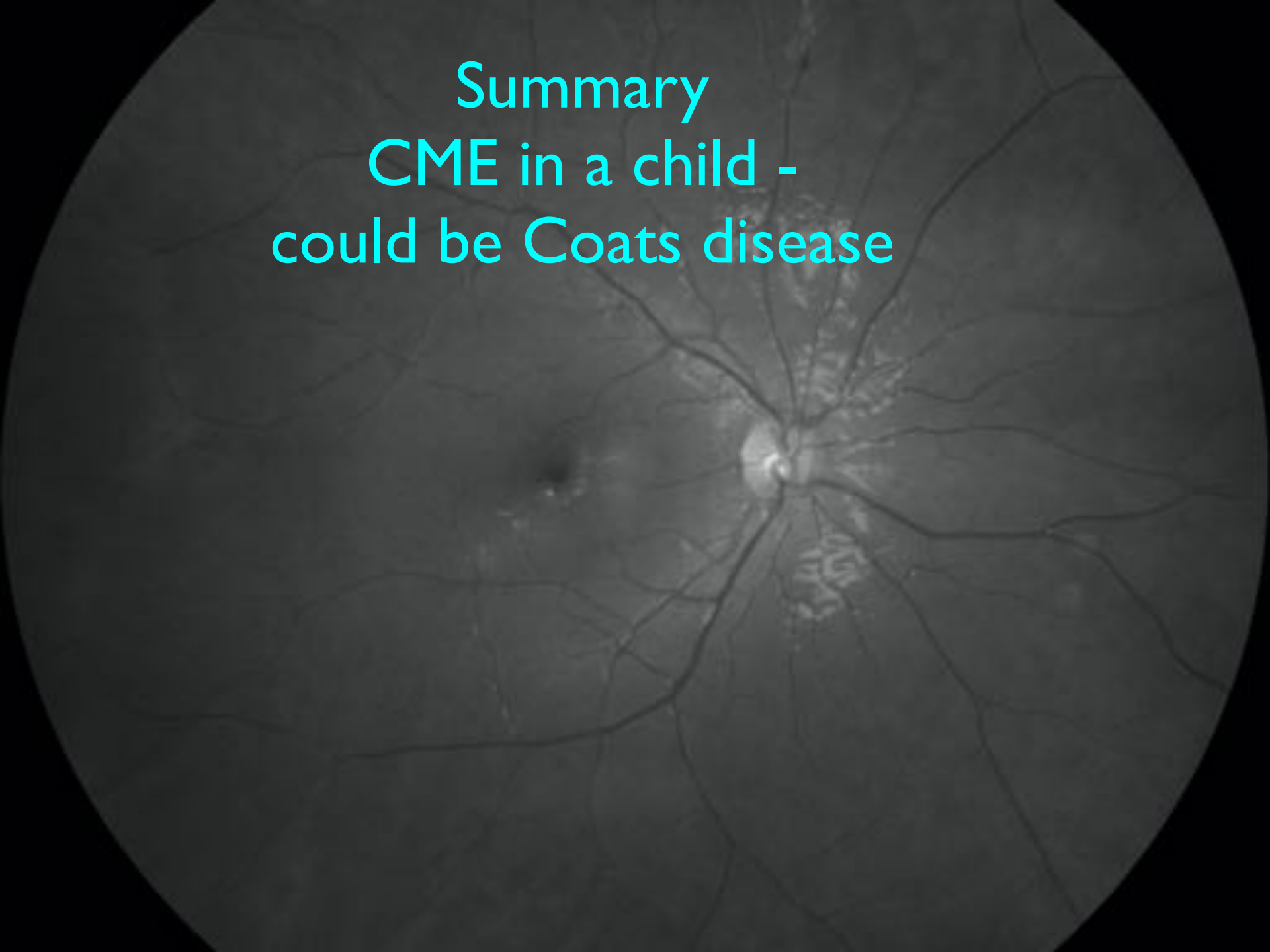
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Financial Disclosure

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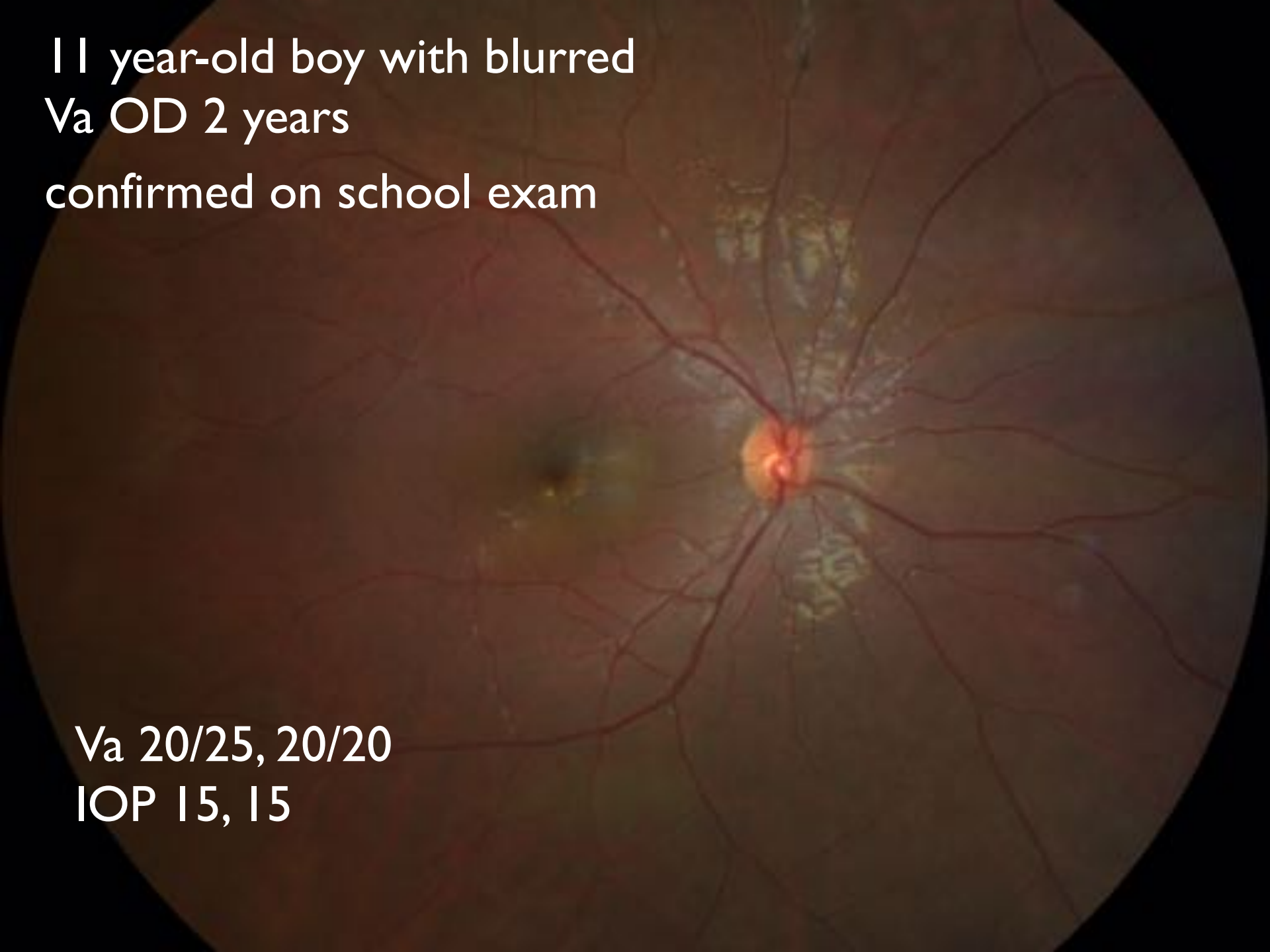
- I have no financial interests or relationships to disclose.

Summary
CME in a child -
could be Coats disease

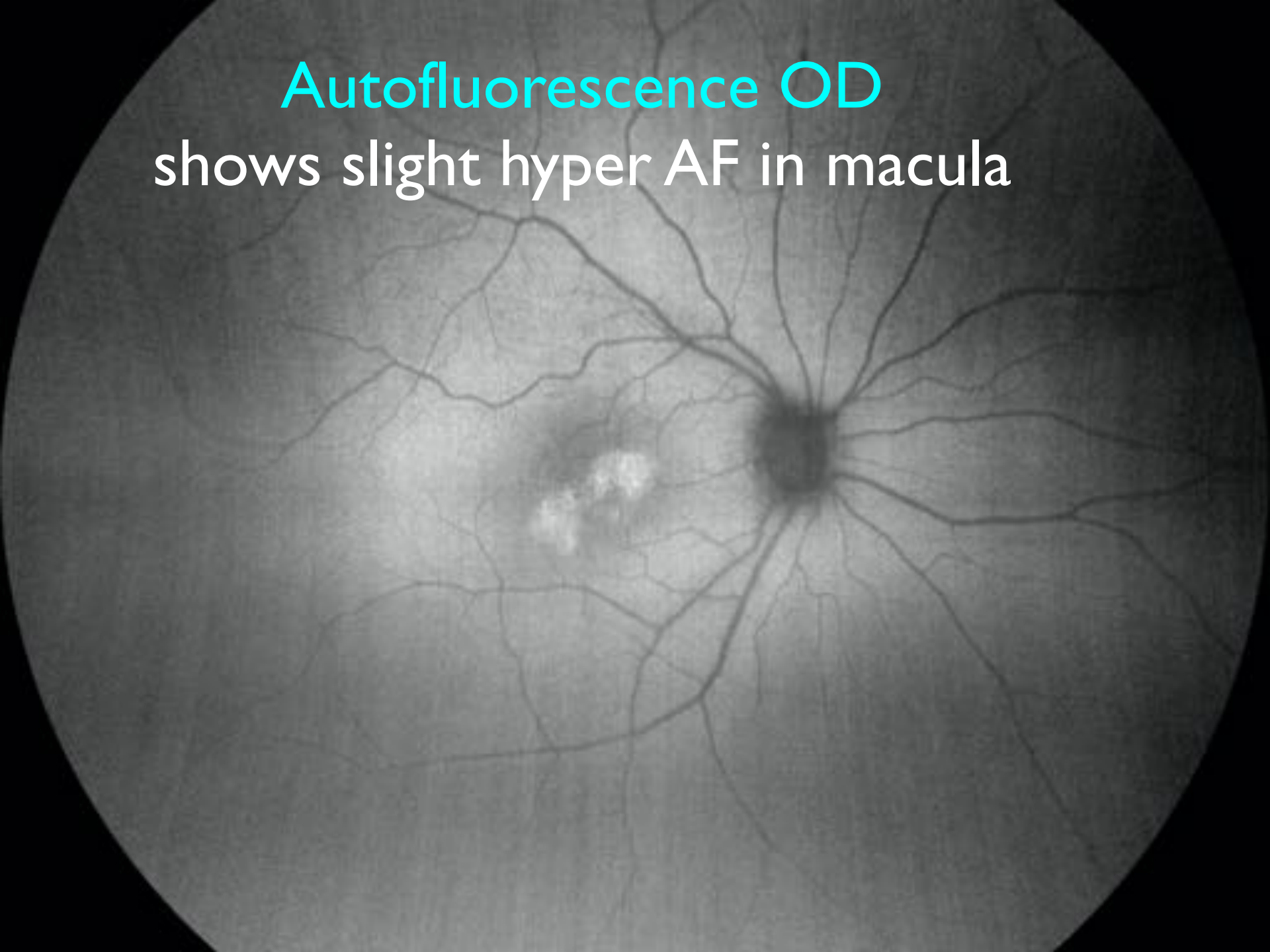


11 year-old boy with blurred
Va OD 2 years
confirmed on school exam

Va 20/25, 20/20
IOP 15, 15

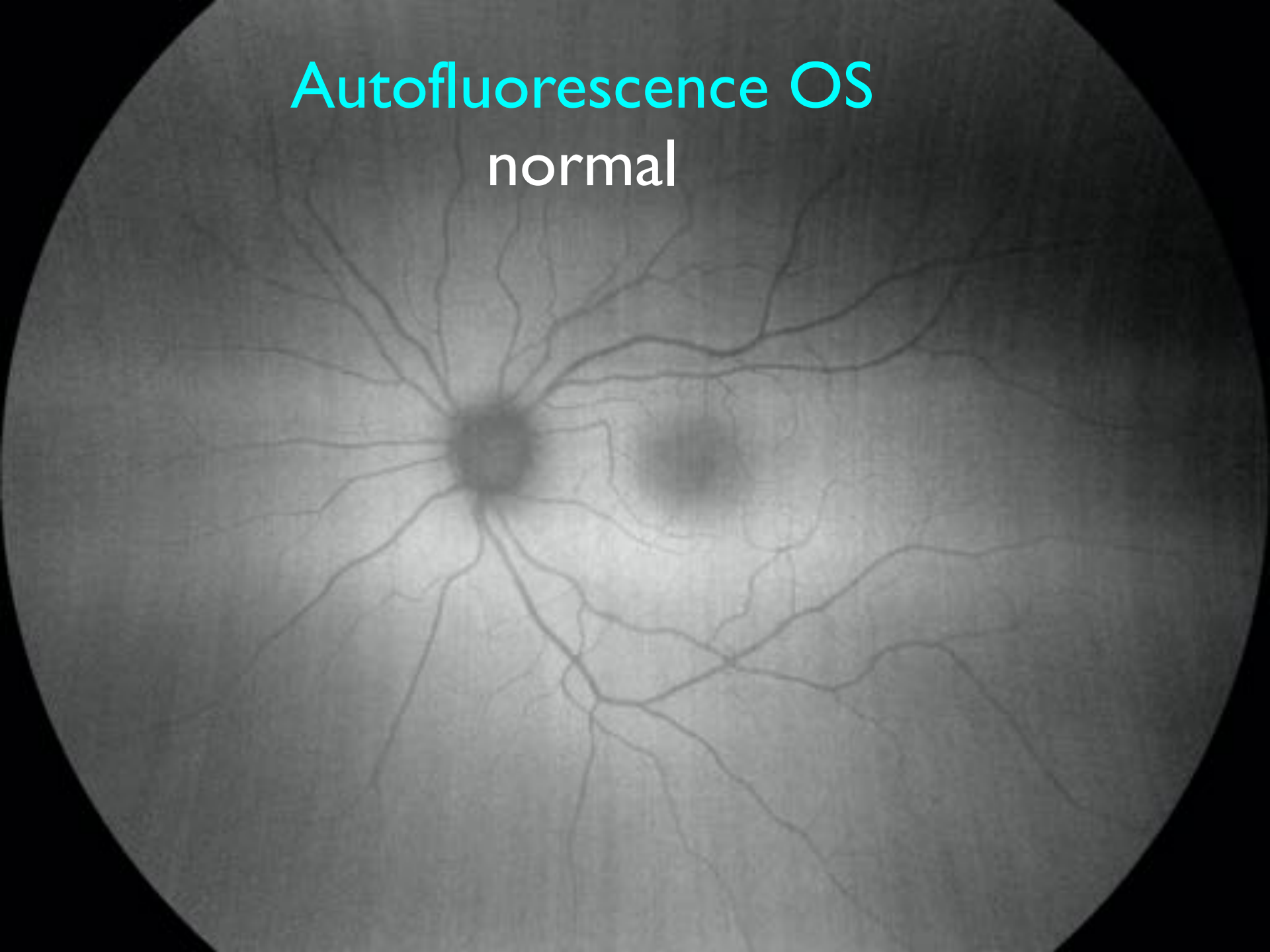


Autofluorescence OD
shows slight hyper AF in macula

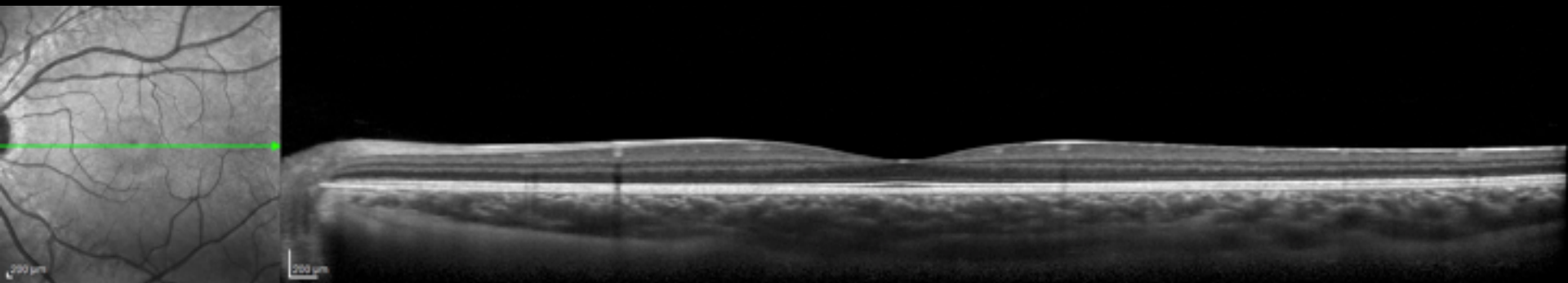


Autofluorescence OS

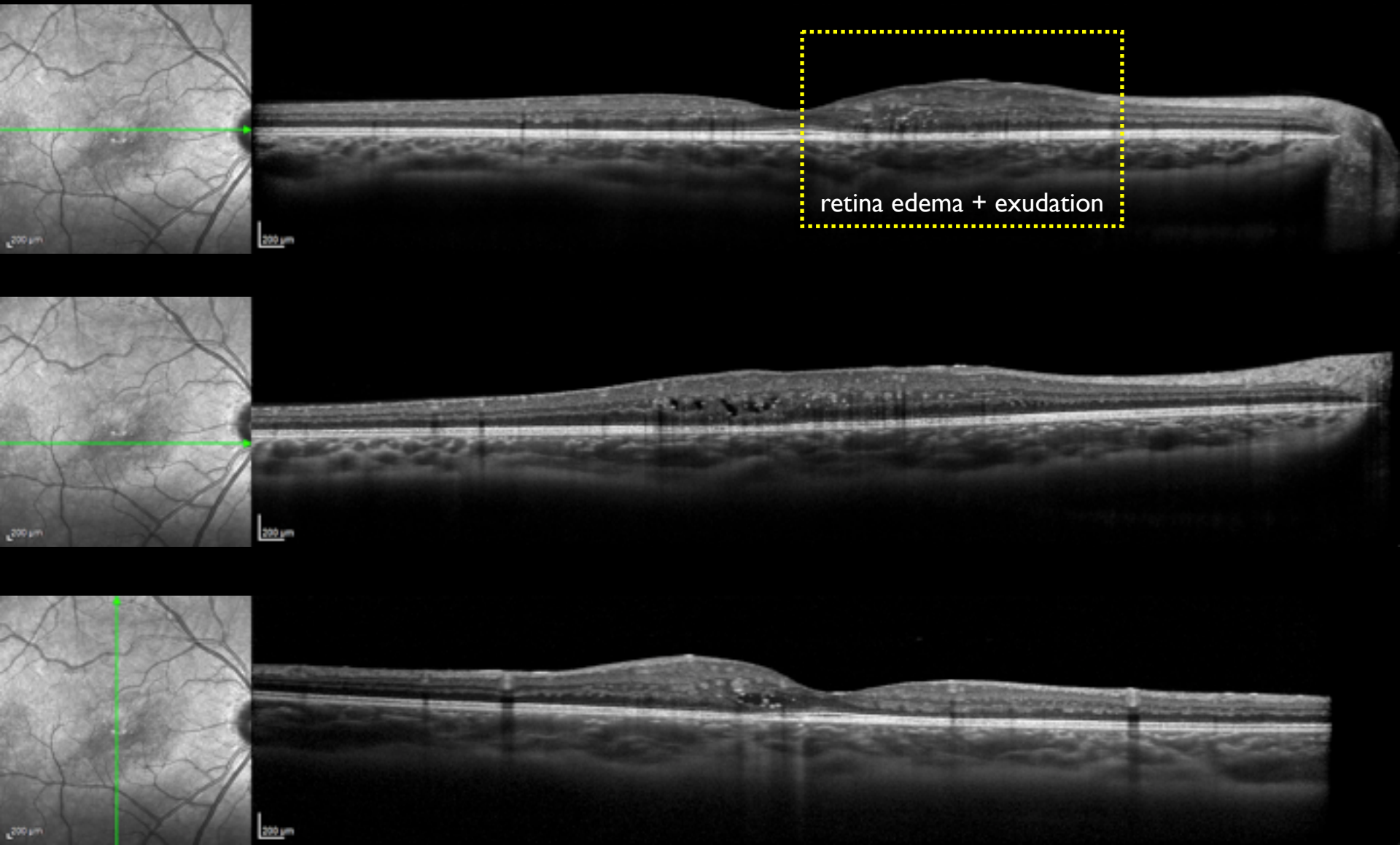
normal



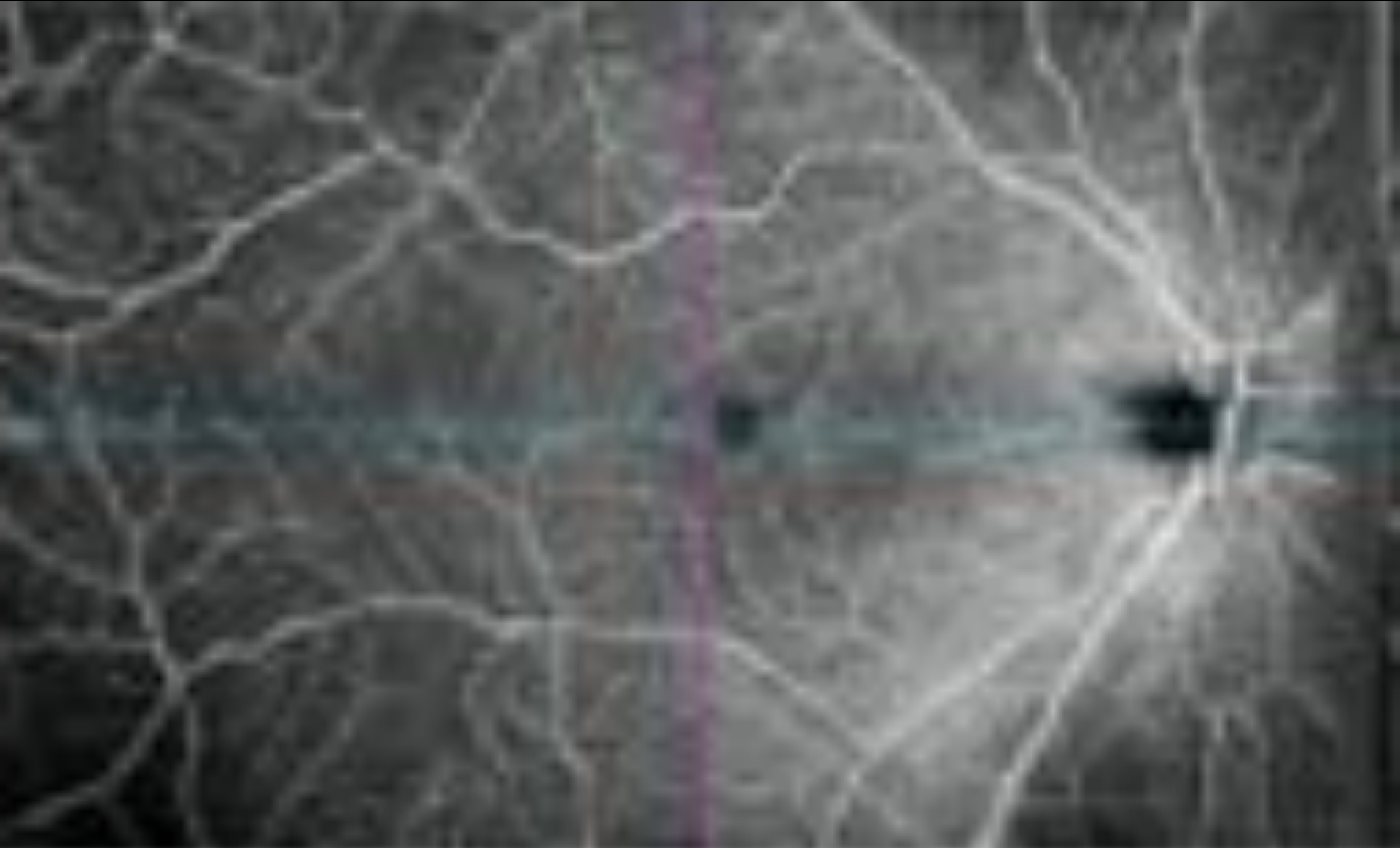
OCT OS



OCT OD

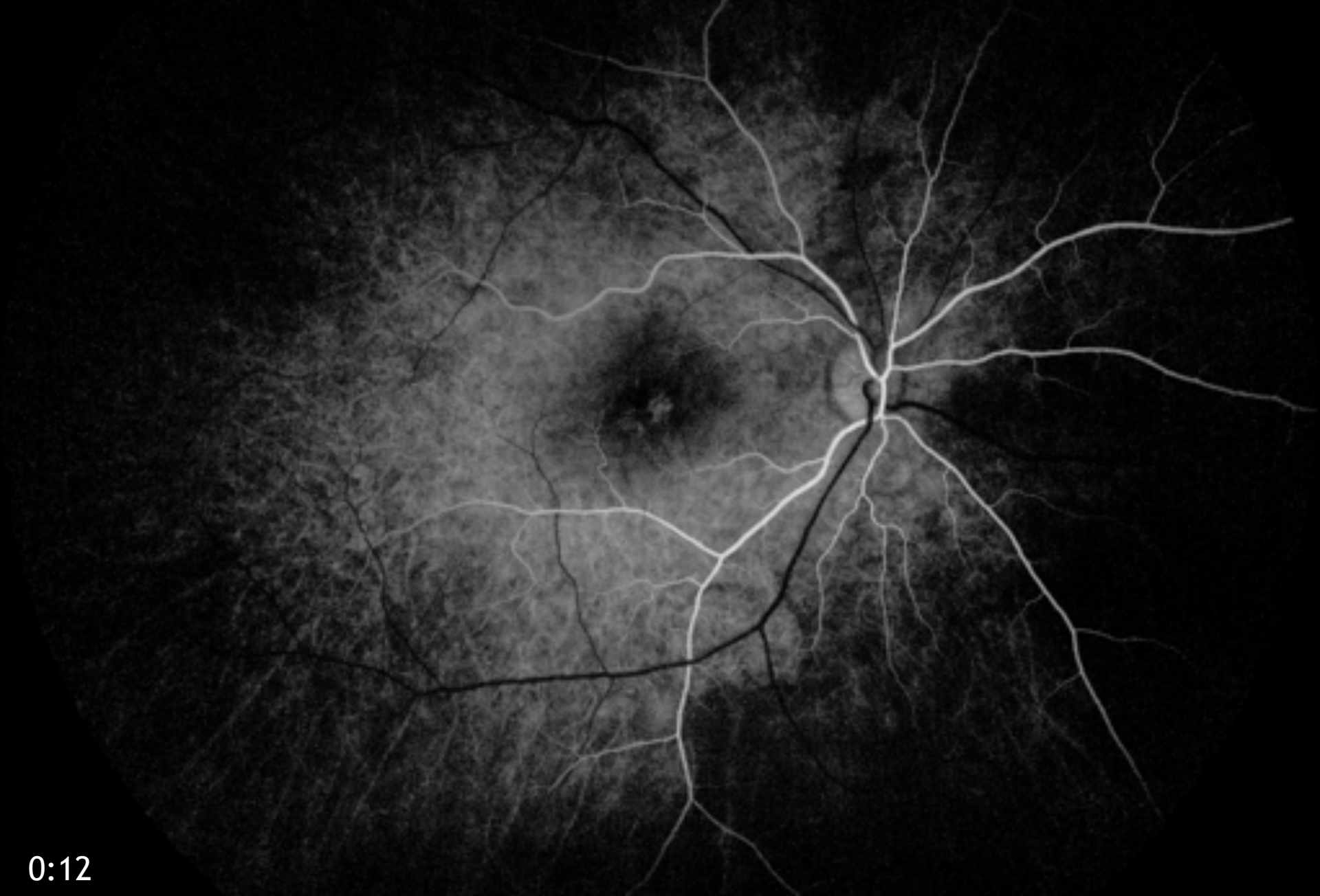


OCTA OD



Slight irregularity of the perifoveal capillaries with
trace reduction in capillary vascular density

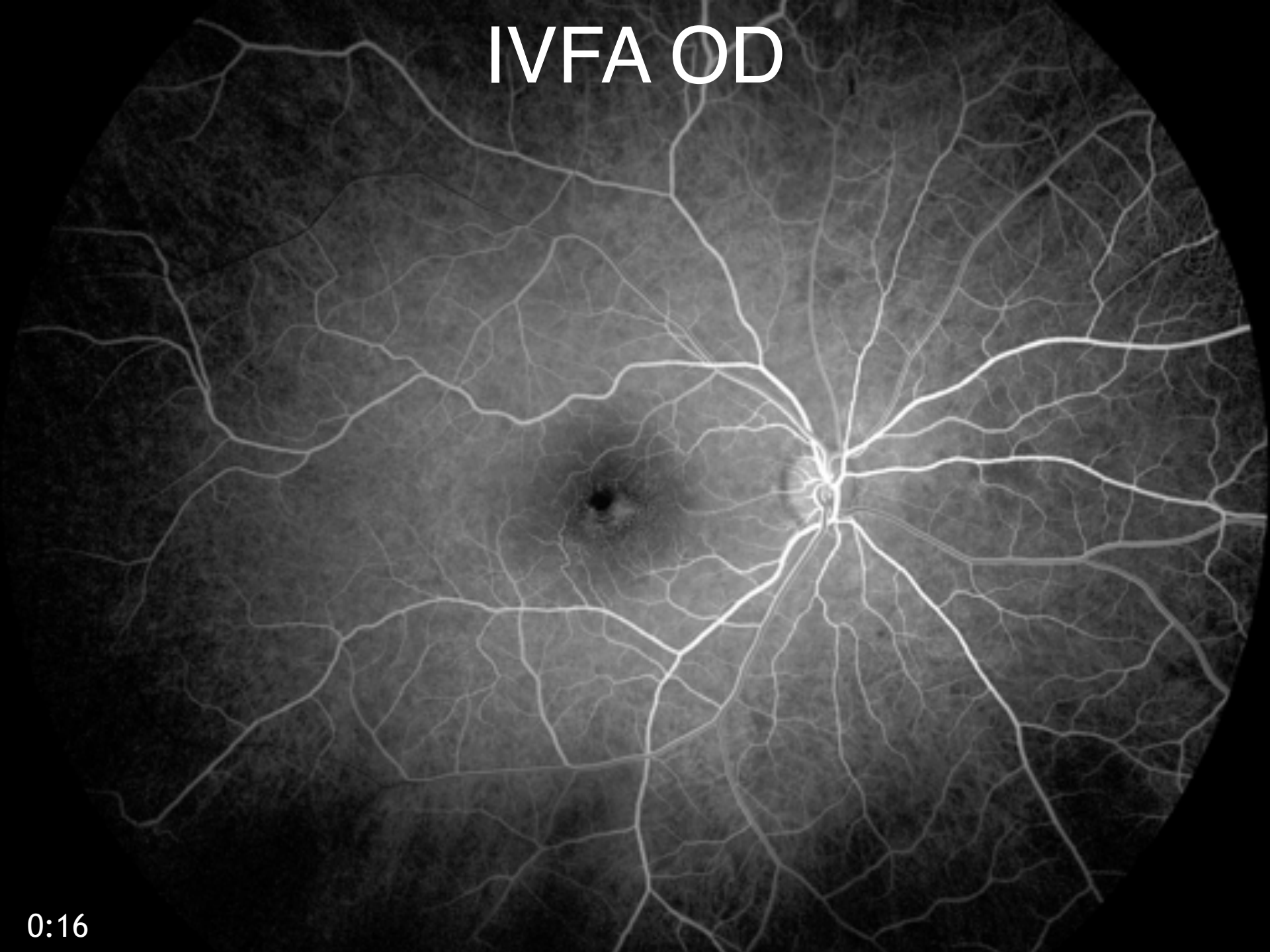
IVFA OD



0:12

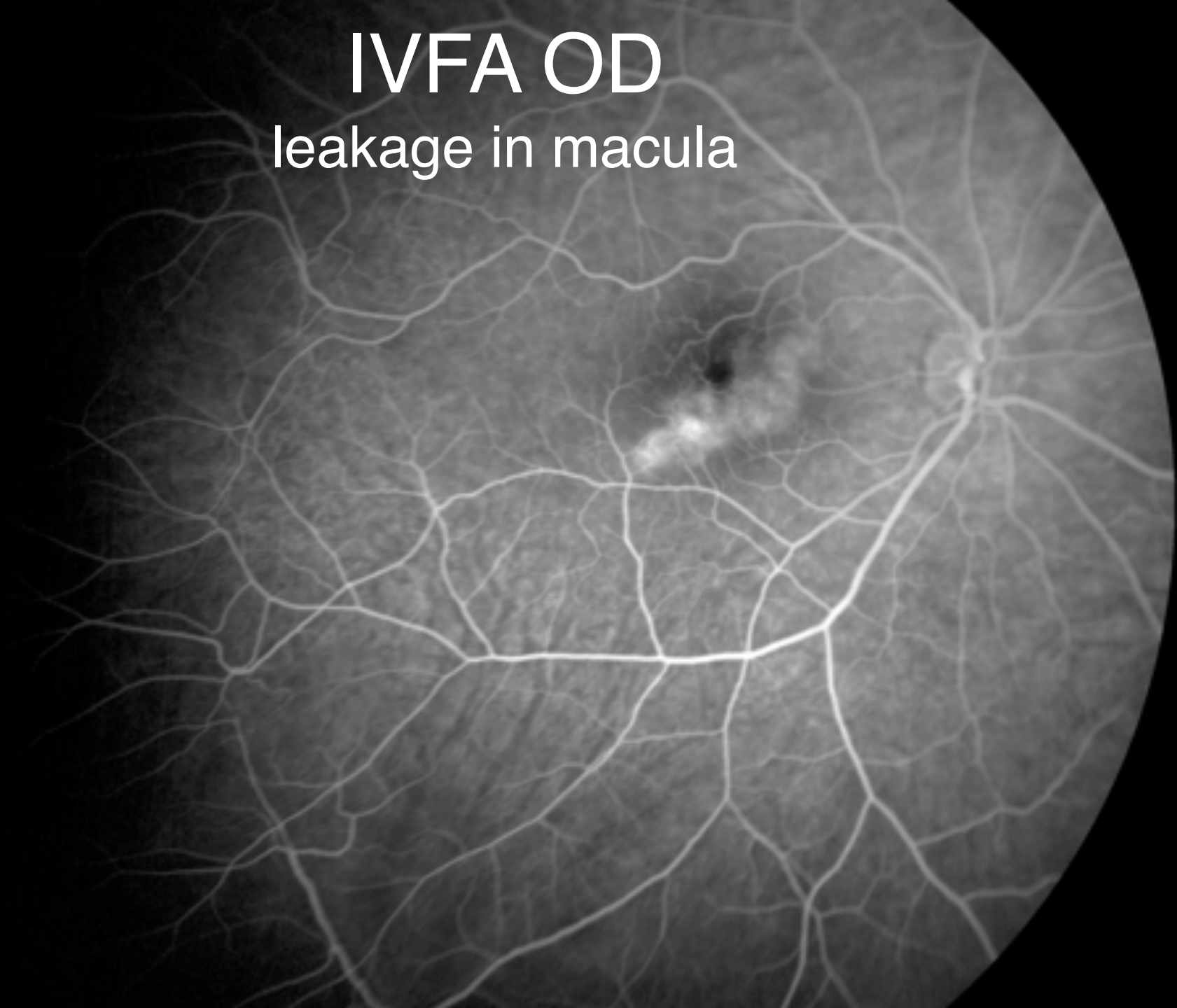
IVFA OD

0:16

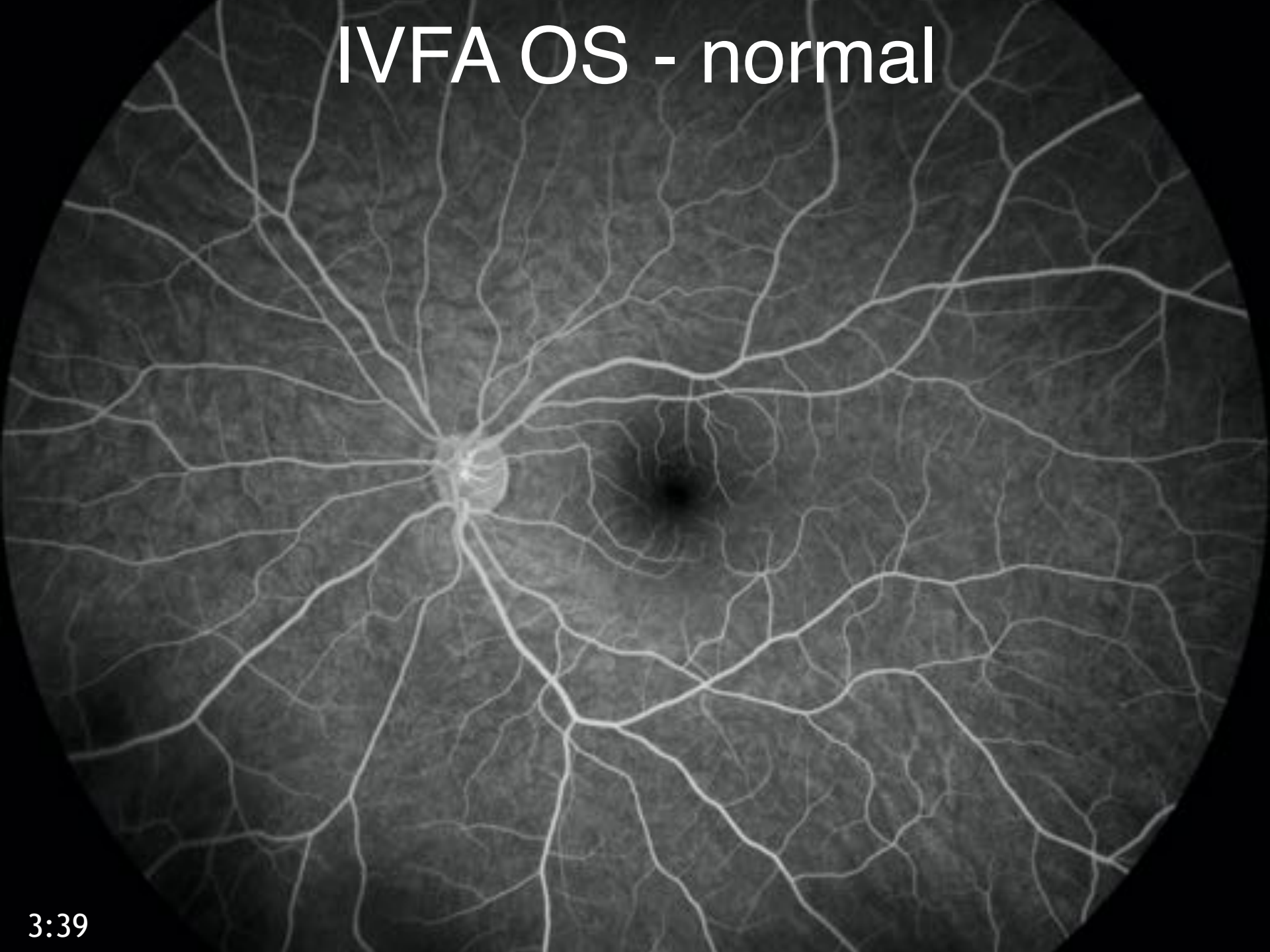


IVFA OD

leakage in macula



IVFA OS - normal



Cystoid Macular Edema in a kid

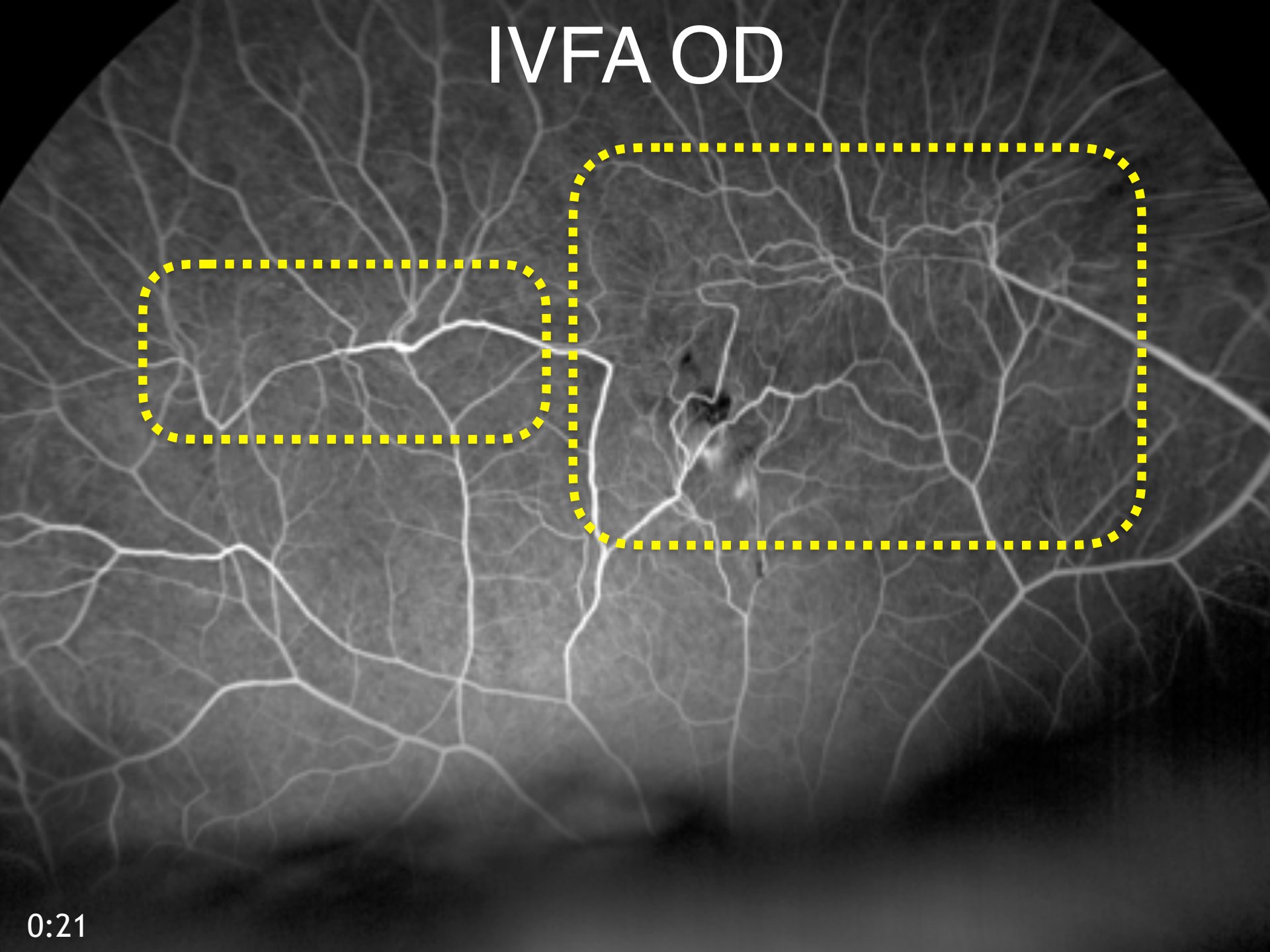
- Congenital
- Vascular
 - Diabetes mellitus
 - BRVO
 - Coats
- Inflammatory
- Medications
- Tumor (VPT, hemangioblastoma, adenoma)
- Trauma

Check the periphery

- Degeneration

Let's look further ...

IVFA OD



IVFA OD



Peripheral non-perfusion, telangiectasia, microaneurysms
cw Coats disease

Peripheral Retinal Vasculopathy

- Coats disease
- FEVR
- Facioscapulohumeral muscular dystrophy
- Dyskeratosis congenita
- Others

What do we know about Coats disease?

- Classification
- Younger age more advanced
- Vision depends on classification

Classification and Management of Coats Disease: The 2000 Proctor Lecture

JERRY A. SHIELDS, MD, CAROL L. SHIELDS, MD,
SANTOSH G. HONAVAR, MD, HAKAN DEMIRCI, MD, AND JACQUELINE CATER, PhD

*** PURPOSE:** To review the methods and results of management in large series of patients with Coats disease, to determine risk factors for poor visual outcome and detachment, and to propose a practical classification of Coats disease.

*** METHODS:** In a retrospective consecutive series in 170 patients, Coats disease was defined as idiopathic retinal telangiectasis with intravitreal or subretinal exudation without appreciable signs of retinal or vitreal traction. We reviewed our experience with management, including

improvement or stability was achieved in 78% of eyes, and final visual acuity was 20/70 or better in 17 eyes (14%), 20/60 to 20/100 in eight (5%), 20/200 to finger counting in 30 (24%), and final vision to no light perception in 49 (46%). Enucleation was ultimately necessary in 20 eyes (18%).

Risk factors predictive of poor visual outcome (20/200 or worse) included postoperative (P = .01), diffuse (P = .05), or superior (P = .04) location of the telangiectatic and exudation, failed resolution of subret-

inal detachment or exudate, and maculopathy were determined using proportional hazards regression models. Based on these observations, a staging classification of Coats disease, applicable to treatment selection and ocular prognosis, is proposed.

*** RESULTS:** In 117 patients (124 eyes) with a mean follow-up of 48 months (range, 6 months to 29 years) primary management was observation in 22 eyes (18%), cryotherapy in 32 (46%), laser photocoagulation in 16 (11%), various methods of retinal detachment surgery in 20 (17%), and enucleation in 14 (11%). Anatomic

retinal detachment and secondary glaucoma; and stage 5, advanced end-stage disease. Poor visual outcome (20/200 or worse) was found in 7% of eyes with stage 1, 15% with stage 2, 74% with stage 3, and 100% of stages 4 and 5 Coats disease. Enucleation was ultimately necessary in 1% of stage 1 and 2, 1% of stage 3, 36% of stage 4, and 0% of stage 5 disease.

*** CONCLUSIONS:** Carefully selected treatment can anatomicall stabilize or improve the eye with Coats disease in 78% of eyes. However, poor visual outcome of 20/200 or worse commonly results. Patients who present with stages 1 to 3 Coats disease have the best visual prognosis, and patients with stages 4 and 5 have a poor visual prognosis. (Am J Ophthalmol. 2004;137:932-940. © 2004 by Elsevier Science Inc. All rights reserved.)

COATS DISEASE IS A NONHEREDITARY CONDITION that is characterized by idiopathic retinal telangiectasis, exudation, and progressive retinal detachment.¹⁻⁴ It usually occurs unilaterally in young males and, if untreated, can lead to total retinal detachment and secondary glaucoma, sometimes requiring enucleation.⁵⁻⁷ There have been no reports that analyze statistically the

Clinical Variations and Complications of Coats Disease in 150 Cases: The 2000 Sanford Gifford Memorial Lecture

JERRY A. SHIELDS, MD, CAROL L. SHIELDS, MD, SANTOSH G. HONAVAR, MD, AND HAKAN DEMIRCI, MD

*** PURPOSE:** The purpose of this report is to review the clinical variations and natural course of Coats disease, using strict diagnostic guidelines.

*** METHODS:** In a retrospective, consecutive series, Coats disease was defined as idiopathic retinal telangiectasis with intravitreal or subretinal exudation without appreciable signs of retinal or vitreal traction. We reviewed our experience with the clinical features, complications, and therapeutic approaches to Coats disease.

was included in the macular area in two eyes (1%), involved nearly the temporal hemis in 66 eyes (42%), inferior hemis in 41 eyes (28%), and more than one sector in 34 eyes (22%). Retinal exudation was present in all 12 clock hours in 66 eyes (33%) and six or more clock hours in 127 eyes (83%). There was a total retinal detachment in 74 eyes (49%) and macular gliosis in 12 (8%). Retinal neovascularization was present in 18 eyes (12%), a secondary glaucoma from the retinal area (5%) and

was the predominant factor in 14 eyes. The most common retinal diagnosis was Coats disease in 146 eyes (97%) and retinoblastoma in 45 (30%). The best response or sign was decreased visual acuity in 66 cases (44%), exudation in 70 (47%), leukocoria in 54 (36%), and 11 patients (8%) were asymptomatic. Visual acuity at presentation was 20/200 or less light perception in 123 eyes (82%). The anterior segment was normal in 147 eyes (98%).

The retinal telangiectasis involved the peripheral or peripheral hemis in 155 of the 150 eyes (99%) and

more in young males, and less than minor visual loss resulting from exudative retinal detachment. The clinician should follow strict criteria in making the diagnosis, to avoid confusing Coats disease with other forms of exudative retinopathy. (Am J Ophthalmol. 2001;131:764-771. © 2001 by Elsevier Science Inc. All rights reserved.)

IN 1908, COATS DESCRIBED AN ENTITY CHARACTERIZED by unilateral retinal vascular abnormalities and retinal exudation, which usually occurred in young males. In 1912, Leber² reported a condition characterized by multiple retinal neovascular associated with retinal degeneration that was usually unilateral in young males. These conditions subsequently came to be known as Coats disease and Leber's atypical atrophy, respectively. Although there had never before, as common, they were often considered to be two different entities in the first half of the twentieth century.

Now,³ as the Seventh Sanford Gifford Lecture at the Chicago Ophthalmological Society in 1971, retinalized exudation between Coats disease and Leber's atypical atrophy and pointed out that they represented a spectrum of

Accepted for publication Jan 15, 2004.

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This work was supported by the Eye Tumor Research Foundation, Philadelphia, Pennsylvania; the Arnold and Muriel Busch Research Foundation, Tampa, FL; the Shurtz Eye Research Foundation, New York, New York; the C. H. H. Foundation, the National Eye Research Foundation, Philadelphia, PA; the National Eye Research Foundation, New York, New York; the H. H. H. Foundation, and the H. H. H. Foundation, New York, New York.

This work was supported by the Eye Tumor Research Foundation, Philadelphia, Pennsylvania; the Arnold and Muriel Busch Research Foundation, Tampa, FL; the Shurtz Eye Research Foundation, New York, New York; the C. H. H. Foundation, the National Eye Research Foundation, Philadelphia, PA; the National Eye Research Foundation, New York, New York; the H. H. H. Foundation, and the H. H. H. Foundation, New York, New York.

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Two early reports on classification

Coats disease classification

Stage 1 Telangiectasia

Stage 2 T Exudation

Stage 3 T E Subretinal fluid

Stage 4 T E S Glaucoma

Stage 5 T E S G Phthisis

Coats disease classification simplified

		<u>150 cases</u>
Stage 1	T	1%
Stage 2	T E	14%
Stage 3	T E S	68%
Stage 4	T E S G	15%
Stage 5	T E S G P	2%

Coats Disease: Clinical Features and Outcomes by Age Category in 351 Cases

Lauren A. Dalvin, MD; Sanika Udyaver, BS; Li-Anne S. Lim, MD; Mehdi Mazloumi, MD, MPH; Hatice T. Atalay, MD; Chloe T. L. Khoo, MD; Carol L. Shields, MD

JPOS 2019

ABSTRACT

Purpose: To investigate features and outcomes of Coats disease by patient age.

Methods: Patients with Coats disease from 1973 to 2018 were evaluated based on age category at presentation

Conclusions: Younger patients (3 years or younger) with Coats disease present with worse visual acuity and more advanced disease stage, and are more likely to require ultimate enucleation.

[*J Pediatr Ophthalmol Strabismus*. 2019;56(5):288-296.]

Regarding Coats disease per age

disease diagnosed (2 vs 6 vs 27 years, $P < .001$). The youngest age group had more referral diagnoses of retinoblastoma (29% vs 15% vs 0%, $P < .001$), worse presenting visual acuity ($< 20/200$: 80% vs 67% vs 31%, $P < .001$), more advanced Coats disease stage (stage 3B: 65% vs 38% vs 10%, $P < .001$), and greater clock-hour extent of telangiectasia (7 vs 5 vs 4, $P < .001$), light bulb aneurysms (7 vs 4 vs 3, $P < .001$), exudation (10 vs 7 vs 5, $P < .001$), and

vascular disorder characterized by retinal telangiectasia, micro and macro “light bulb” aneurysms, and intraretinal and subretinal exudation.¹ Common presenting features include vision loss, strabismus, and xanthocoria, the latter of which can mimic leukocoria of retinoblastoma.¹ Coats disease is classified into five stages of increasing disease severity, ranging from asymptomatic retinal telangiectasia (stage 1) to

Coats disease based on age

- Dalvin et al 2019
- n=351 pts

Outcomes

- (≤ 3 vs. $>3-10$ vs. >10 years)
 - worse final Va $<20/200$ (83% vs. 64% vs. 39%, $p<0.001$)
 - require enucleation (22% vs 10% vs 6%, $p=0.010$)

Visual acuity outcomes in Coats disease by classification stage in 160 patients

British J Ophthalmol 2019

Carol L Shields,^{*} Sanika Udyaver, Lauren A Dalvin, Li-Anne S Lim, Hatice T Atalay, Chloe Khoo, Mehdi Mazloumi, Jerry A Shields

ABSTRACT

Purpose To assess visual outcomes of Coats disease by classification stage

at initial visit and indicated that vision outcome was '*dismal*', including 20/200–20/400 (n=2), counting fingers (n=2), light perception (n=1)

In 2000, Char reviewed 10 patients with Coats disease with mean of 2.4 years at initial visit and indicated that vision outcome was '*dismal*', including 20/200–20/400 (n=2), counting fingers (n=2), light perception (n=1) and no light perception (n=5).⁴

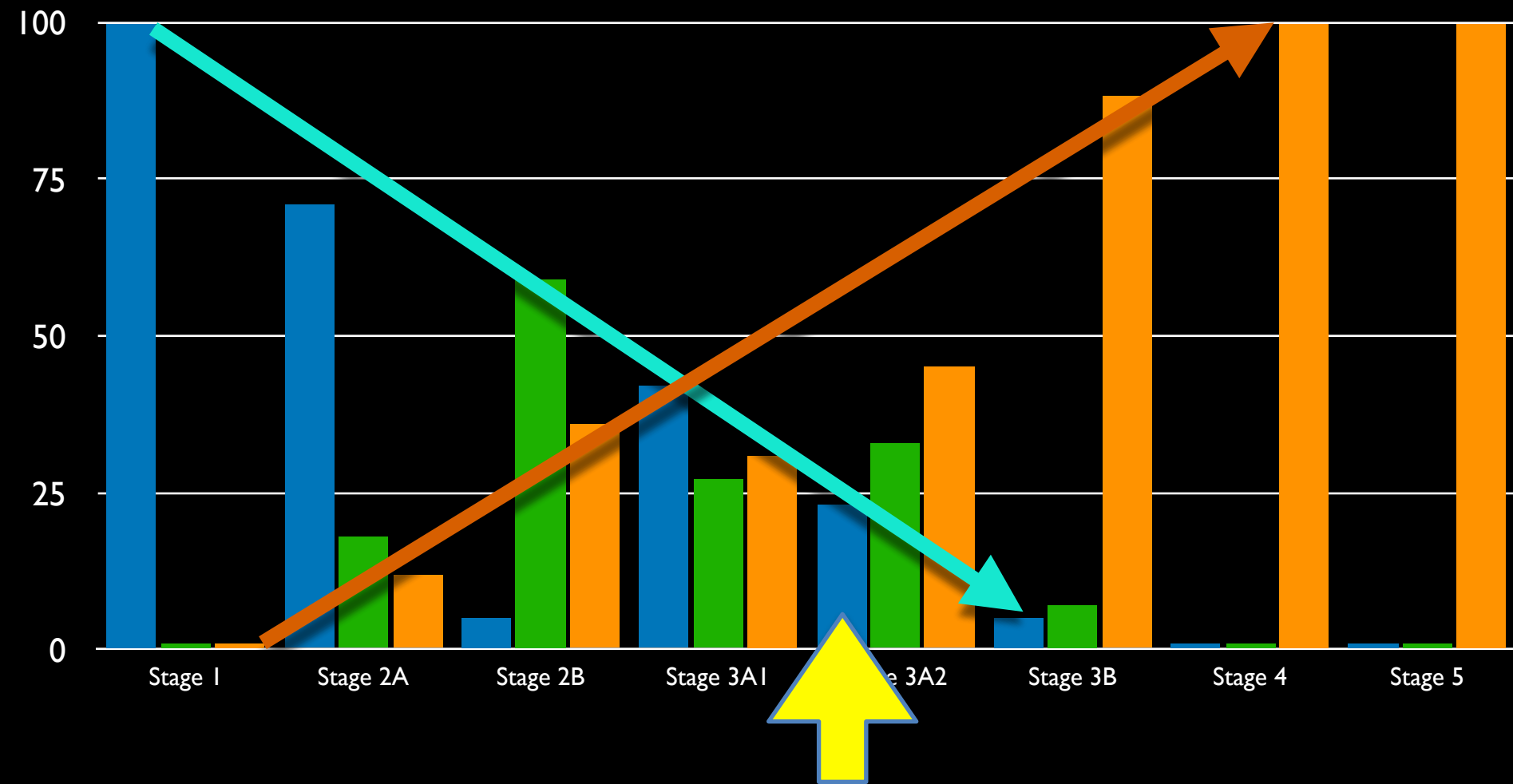
Coats disease: Visual outcome

20/20-20/40

20/50-20/200

20/400-NLP

$p < 0.001$



... back to our case





Summary

- 11 yowm
- presented with CME
- Coats disease
- treated laser
- anticipate fair visual outcome