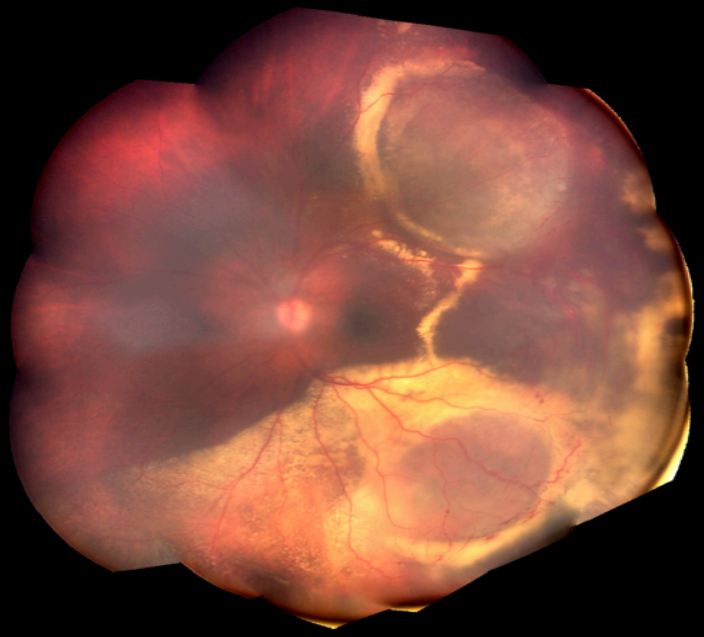
Retinoschisis in Coats Disease: clinical picture, therapeutic considerations and management outcomes



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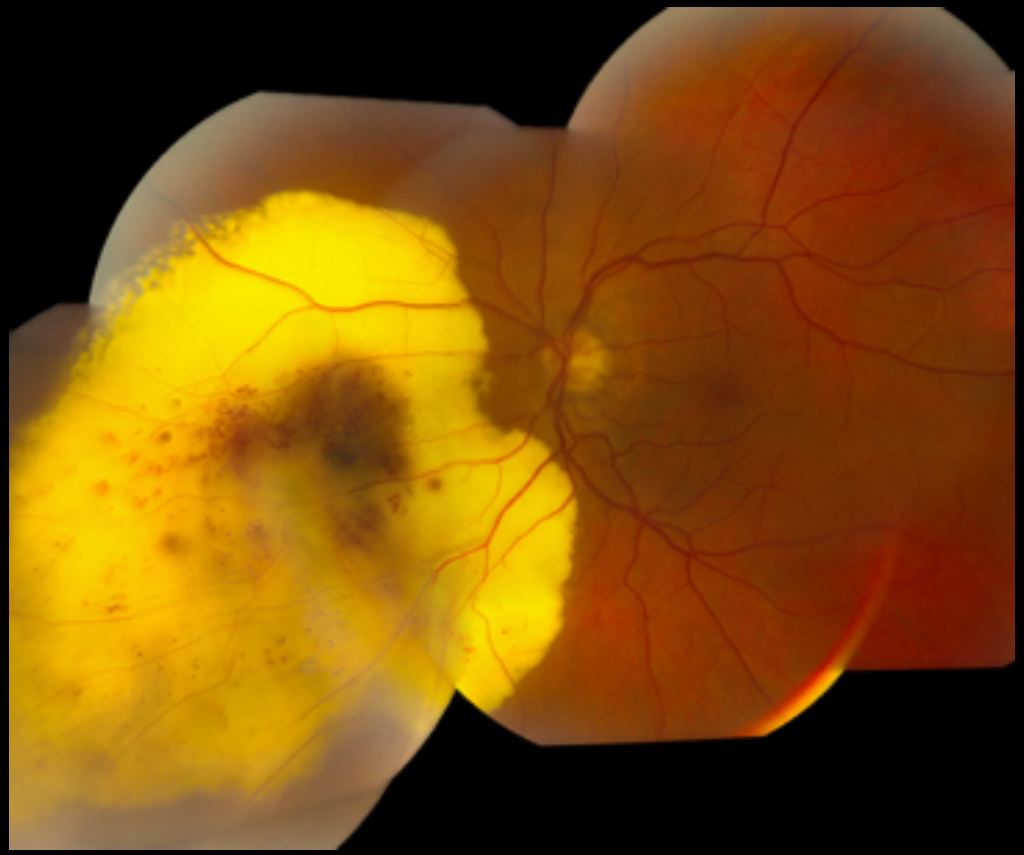
No relevant disclosures for this presentation

ALCON, DORC, VISUNEX, ZEISS, PHOENIX, ALLERGAN

Aerie, ProQR, AGTC

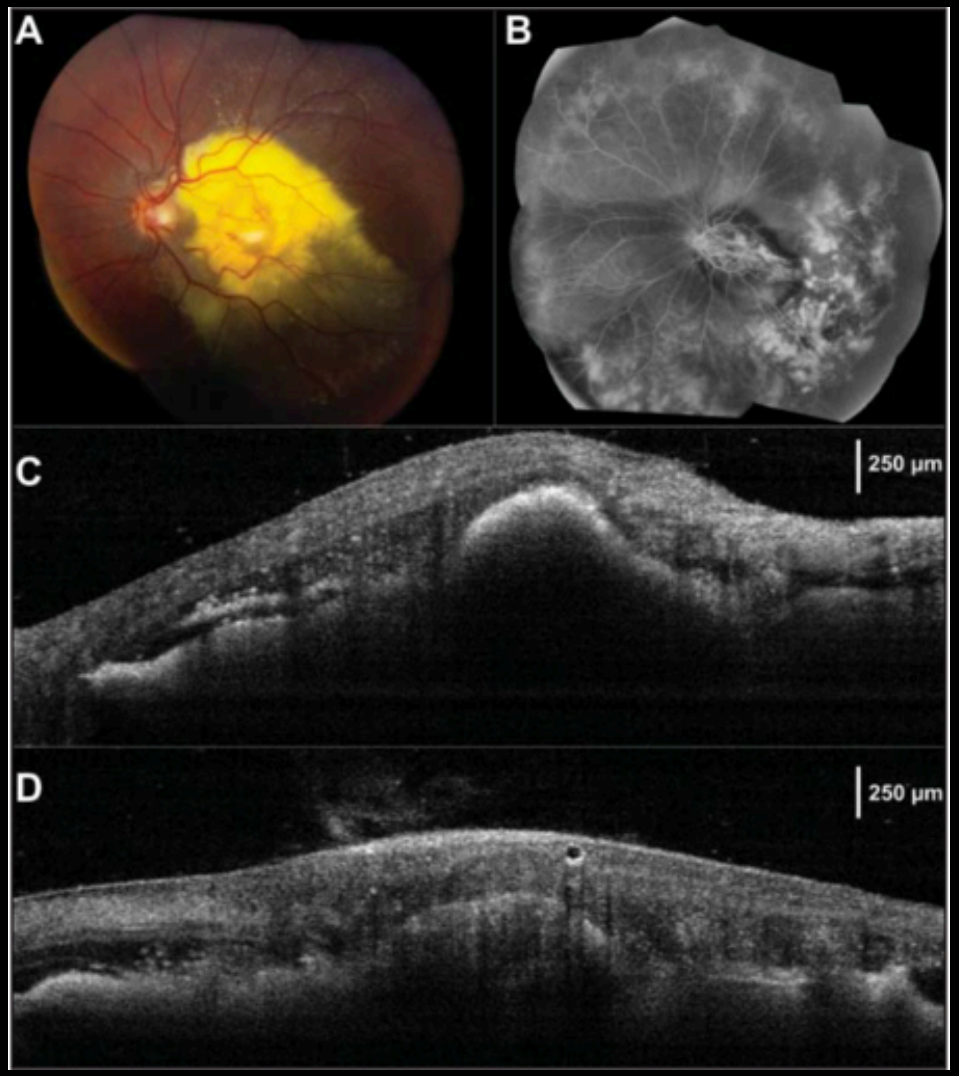
Coats Disease: Background



- Congenital, Non-hereditary
- Unilateral
- Males > Females
- Presenting Symptom:
  - Decreased VA (43%)
  - Strabismus (37%)
  - Leukocoria (20%)

Ong SS, Buckley EG, McCuen II BW, Jaffe GJ, Postel EA, Mahmoud TH, Stinnett SS, Toth CA, Vajzovic L, Mruthyunjaya P. Comparison of visual outcomes in Coats' disease: A 20-year experience. Ophthalmology 2017;-:1e9 a 2017
Shields et al. Clinical variations and complications of coats disease in 150 cases: the Sanford Gifford memorial lecture. Am J Ophthalmol.2001;131(5):561-71

Coats Disease: Diagnosis/Monitoring



- Clinical Diagnosis: angiography and ultrasound
- Long-Term Multimodality Imaging
  - *Only 41% of patient present with correct diagnosis*
  - *27% referred for Retinoblastoma*
    - *Shields et al. AJO 2001*

Henry CR, Berrocal AM, Hess DJ, Murray TG. Intraoperative spectral domain optical coherence tomography in Coats disease. OSLI Retina. 2012.

Coats Disease: Globe salvage

ADVANCED COATS' DISEASE

Management With Repetitive Aggressive Laser Ablation Therapy

AMY C. SCHEFLER, MD, AUDINA M. BERROCAL, MD,
TIMOTHY G. MURRAY, MD, MBA

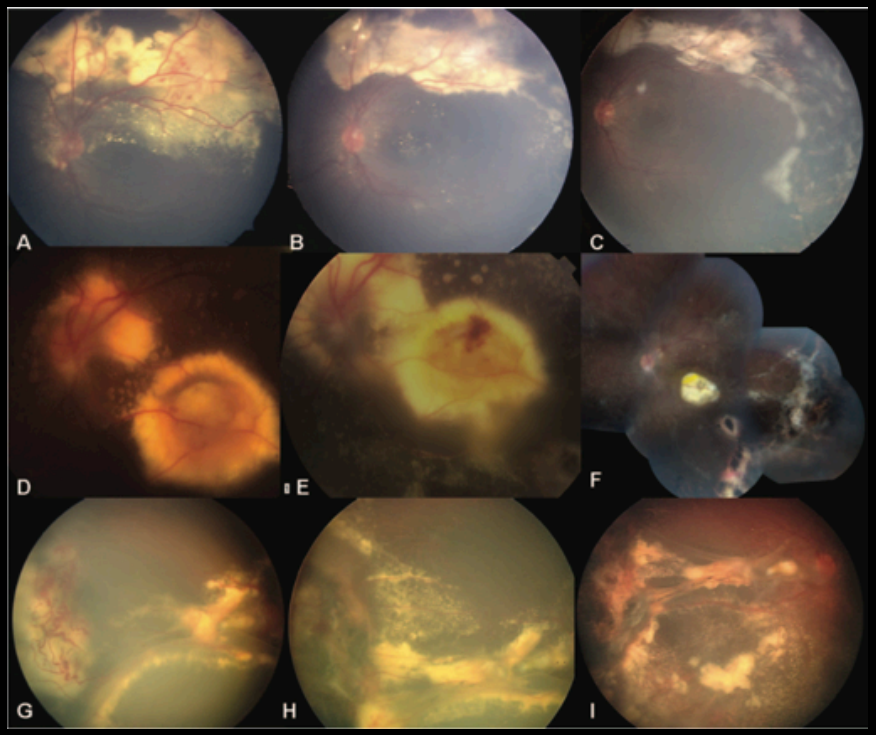


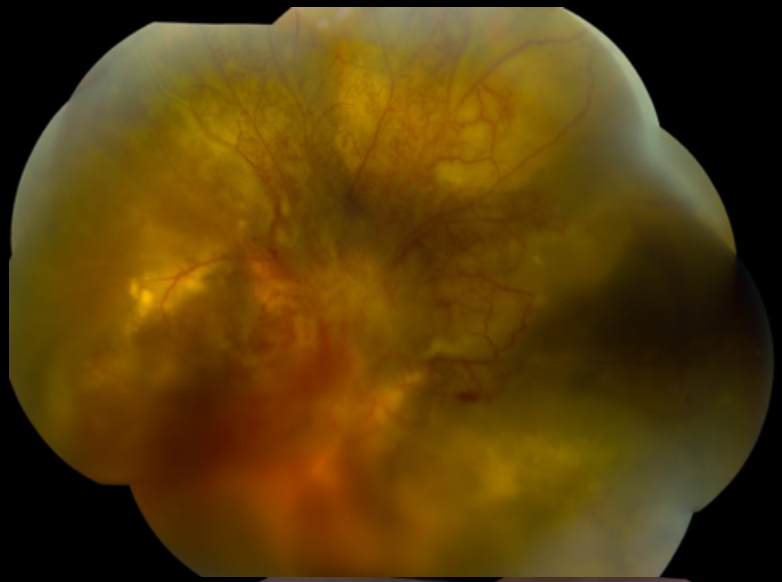
Table 1. Clinical Characteristics of Patients With Coats' Disease

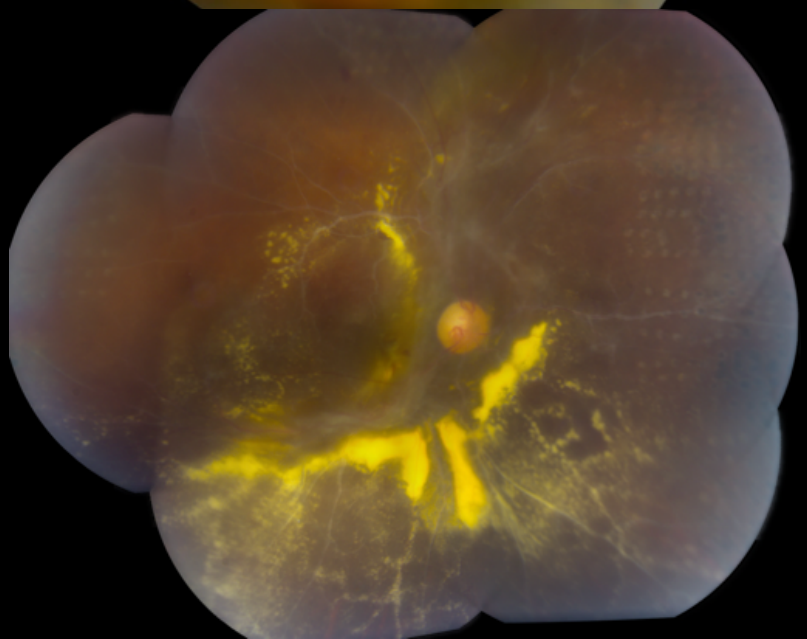
Patient	Sex	R/L	Age at Presentation	Presenting Stage of Disease*	No. of Laser Applications Patient Received	Final Visual Acuity	Outcome	Follow-up (mo)
1	M	L	12 yr 3 mo	2A	5	20/20	Anatomic success	7
2	M	L	9 yr 6 mo	2B	3	20/25	Anatomic success	3
3	M	R	12 yr 8 mo	2B	6	20/20	Anatomic success	14
4	M	R	9 yr 10 mo	3A1	3	20/20	Active exudation currently	3
5	M	R	7 yr 8 mo	3A1	5	20/25	Anatomic success	8
6	M	R	2 yr 5 mo	3A1	7	5/200	Anatomic success	78
7	M	R	1 yr 3 mo	3A1	8	N/A†	Anatomic success	12
8	M	R	10 yr 3 mo	3A1	6	N/A‡	Anatomic success	54
9	M	L	9 yr 6 mo	3A1	3	20/20	Anatomic success	7
10	M	R	9 yr 6 mo	3A2	3	20/80	Anatomic success	6
11	M	R	4 yr 3 mo	3A2	5	5/200	Anatomic success	16
12	M	L	10 yr 1 mo	3B	5	20/25	Anatomic success	33
13	M	L	9 yr 2 mo	3B	9	20/200	Anatomic success	47
14	M	R	16 yr 8 mo	3B	5	LP	Anatomic success	8
15	M	R	2 yr 2 mo	3B	5	LP	Anatomic success	36
16	M	R	5 yr 0 mo	3B	3	NLP	Advanced end-stage disease; non-painful	10 10
17	M	L	2 yr 5 mo	4	2	NLP	Enucleation	9

- Globe Salvation rate 94%
- Visual Acuity 20/20-20/50 in 50% of patients

Schefler AC, Berrocal AM, Murray TG. Adanced coats disease management with repetitive aggressive ablation therapy. Retina. 2008, 28(3).

Coats Disease: Current advance treatment





Treatment of Coats' Disease With Combination Therapy of Intravitreal Bevacizumab, Laser Photocoagulation, and Sub-Tenon Corticosteroids

Julia Sein, MD; Jonathan H. Tzu, MD; Timothy G. Murray, MD; Audina M. Berrocal, MD

August 2002 – January 2014 (26 eyes)

Primary Outcome: Anatomic Success, Globe Salvation, Final Visual Acuity

Median Laser	5 sessions
Mean IVB	3 injections
Globe Salvage	100%

Coats Disease:Macrocyst/schisis

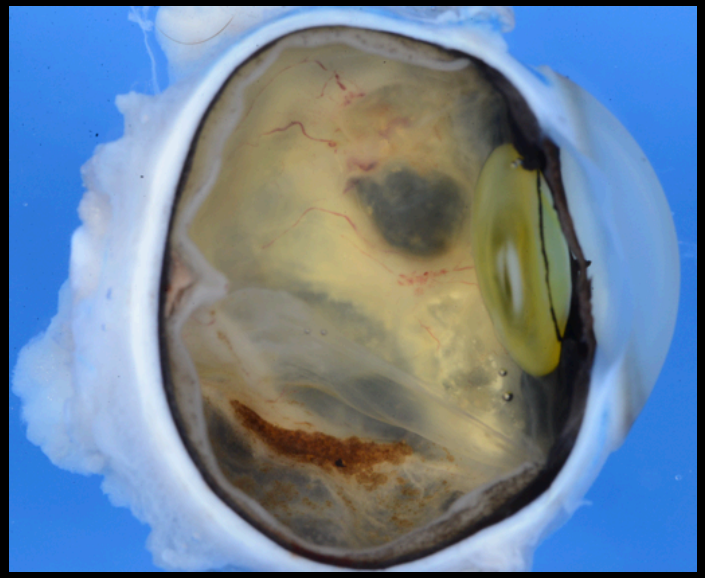
Clinical Variations and Complications of Coats Disease in 150 Cases: The 2000 Sanford Gifford Memorial Lecture

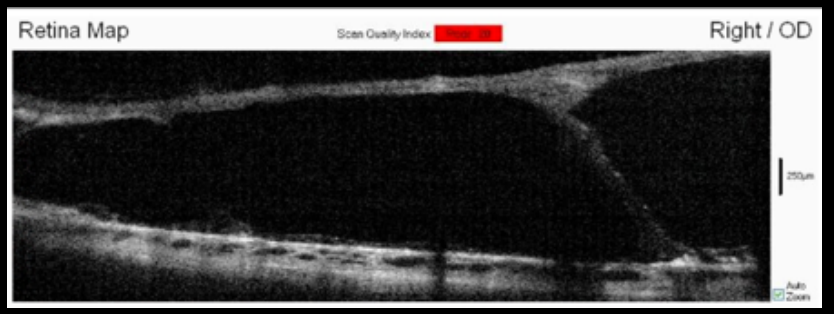
JERRY A. SHIELDS, MD, CAROL L. SHIELDS, MD, SONTASH G. HONAVAR. MD. AND HAKAN DEMIRCI,

Posterior Segment Finding	Number (%)*
Telangiectasia	158 (100%)
Intraretinal exudation	157 (99%)
Exudative retinal detachment	128 (81%)
None	30 (19%)
Partial	54 (34%)
Total	74 (47%)
Retinal hemorrhage	20 (13%)
Retinal macrocyst	18 (11%)
Vasoproliferative tumor	9 (6%)
Optic disk neovascularization	3 (2%)
Retinal neovascularization	1 (1%)
Vitreous hemorrhage	0 (0%)

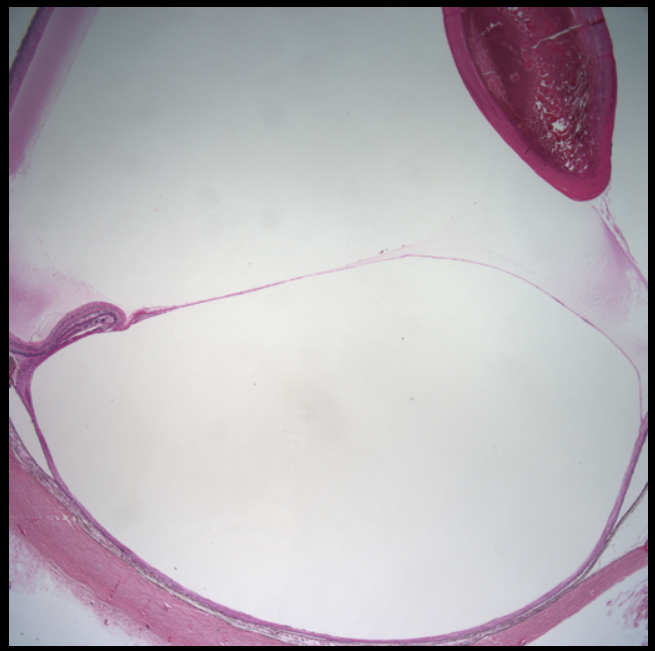
Ong SS, Buckley EG, McCuen II BW, Jaffe GJ, Postel EA, Mahmoud TH, Stinnett SS, Toth CA, Vajzovic L, Mruthyunjaya P. Comparison of visual outcomes in Coats' disease: A 20-year experience. Ophthalmology 2017;-:1e9 a 2017

Coats: Pathology

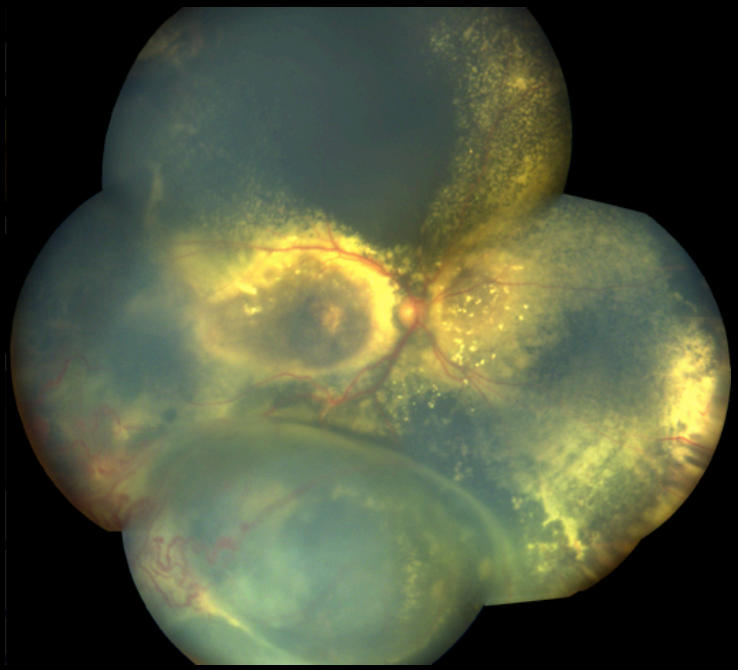


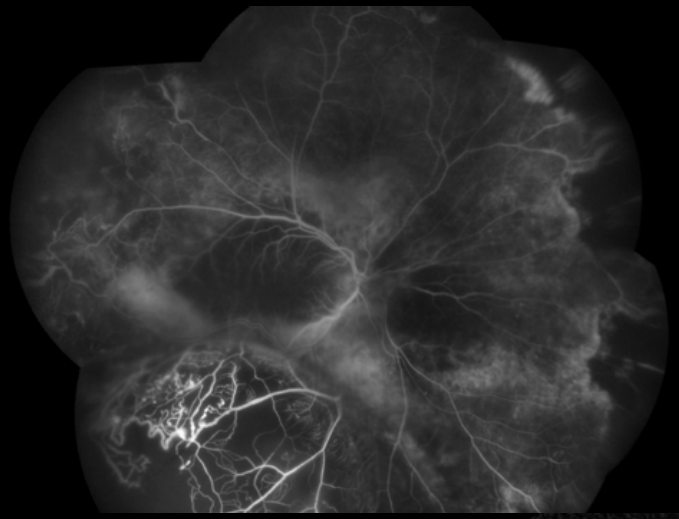


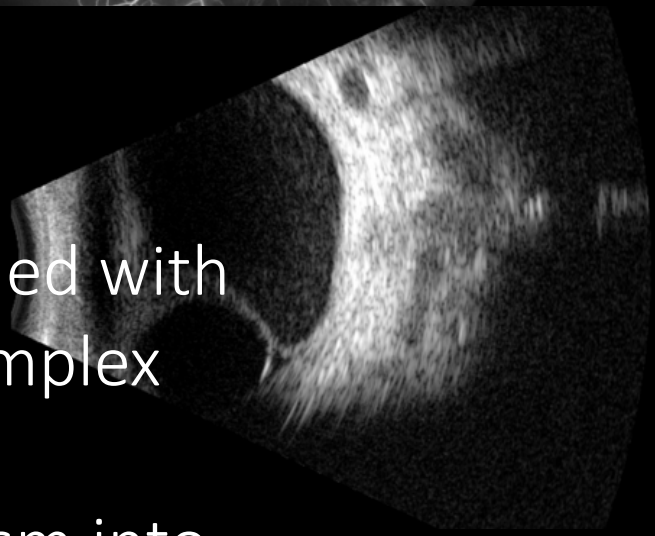
- Splitting of layers in the neurosensory retina
  - Tractional
  - Exudative



Acquired Schisis: Coats Disease

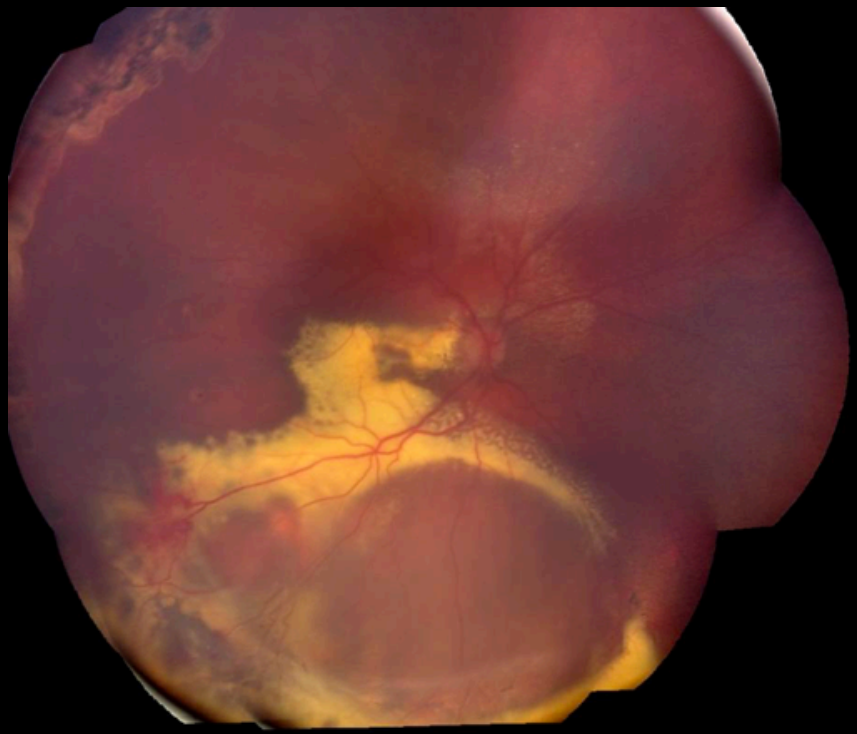






- Tractional: Tractional forces combined with vascular compromise leading to complex schisis
- Exudative: Leakage of microaneurysm into the retinal layers leading to schisis
- Ischemic

Coats Disease: Retinoschisis



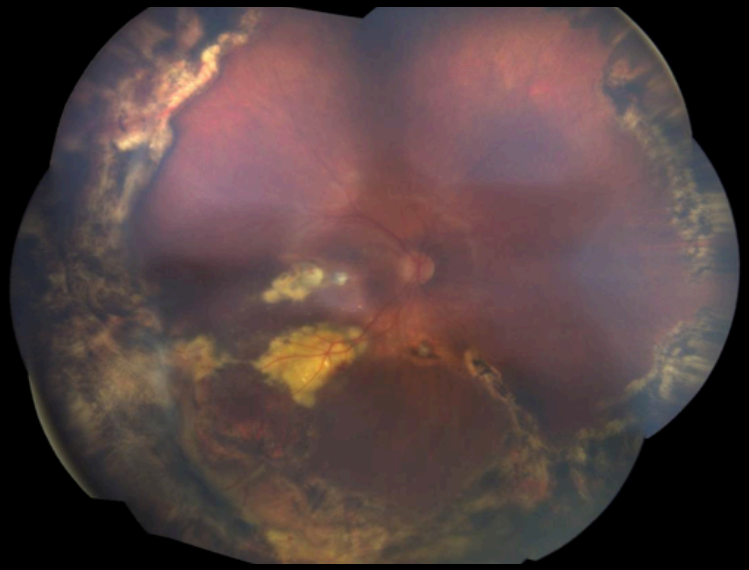
Cavity collapsed with treatment

14 months later:

7 IVFA guided laser treatments

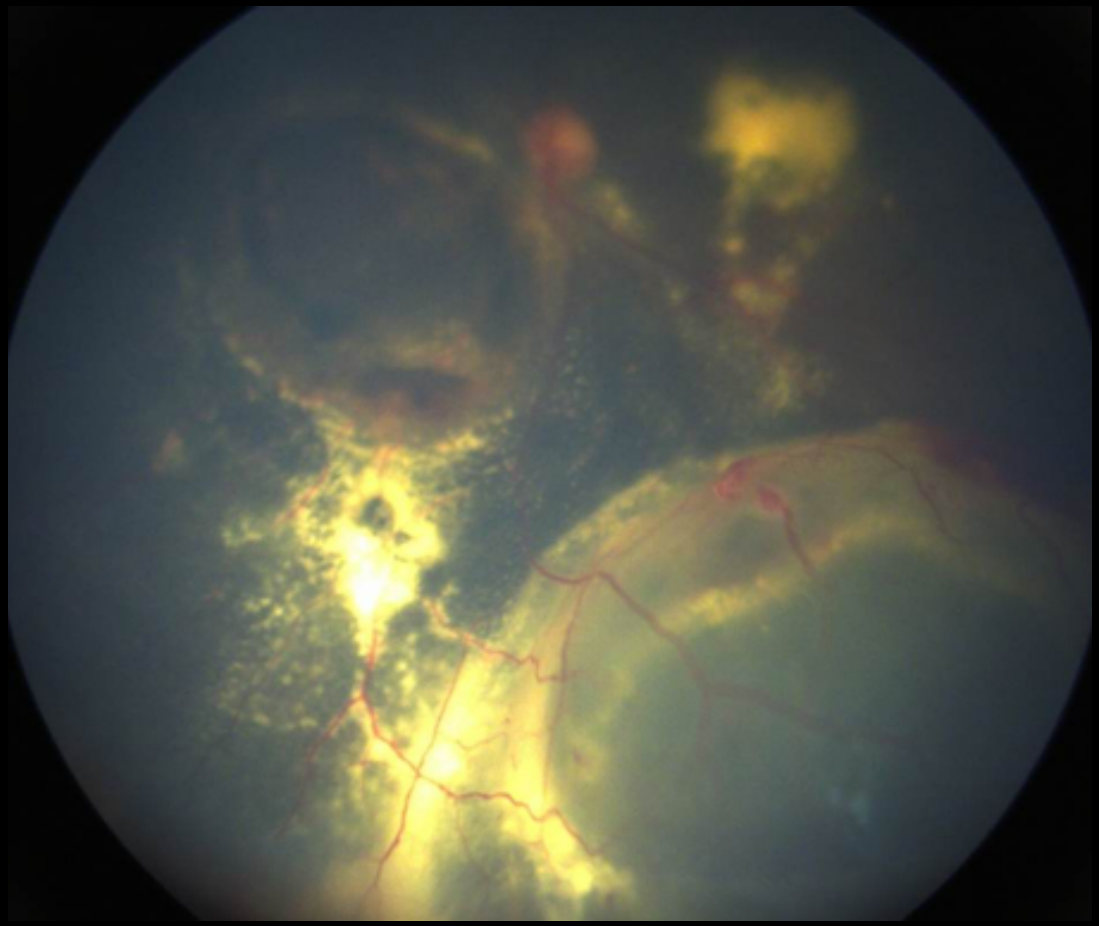
7 IVB

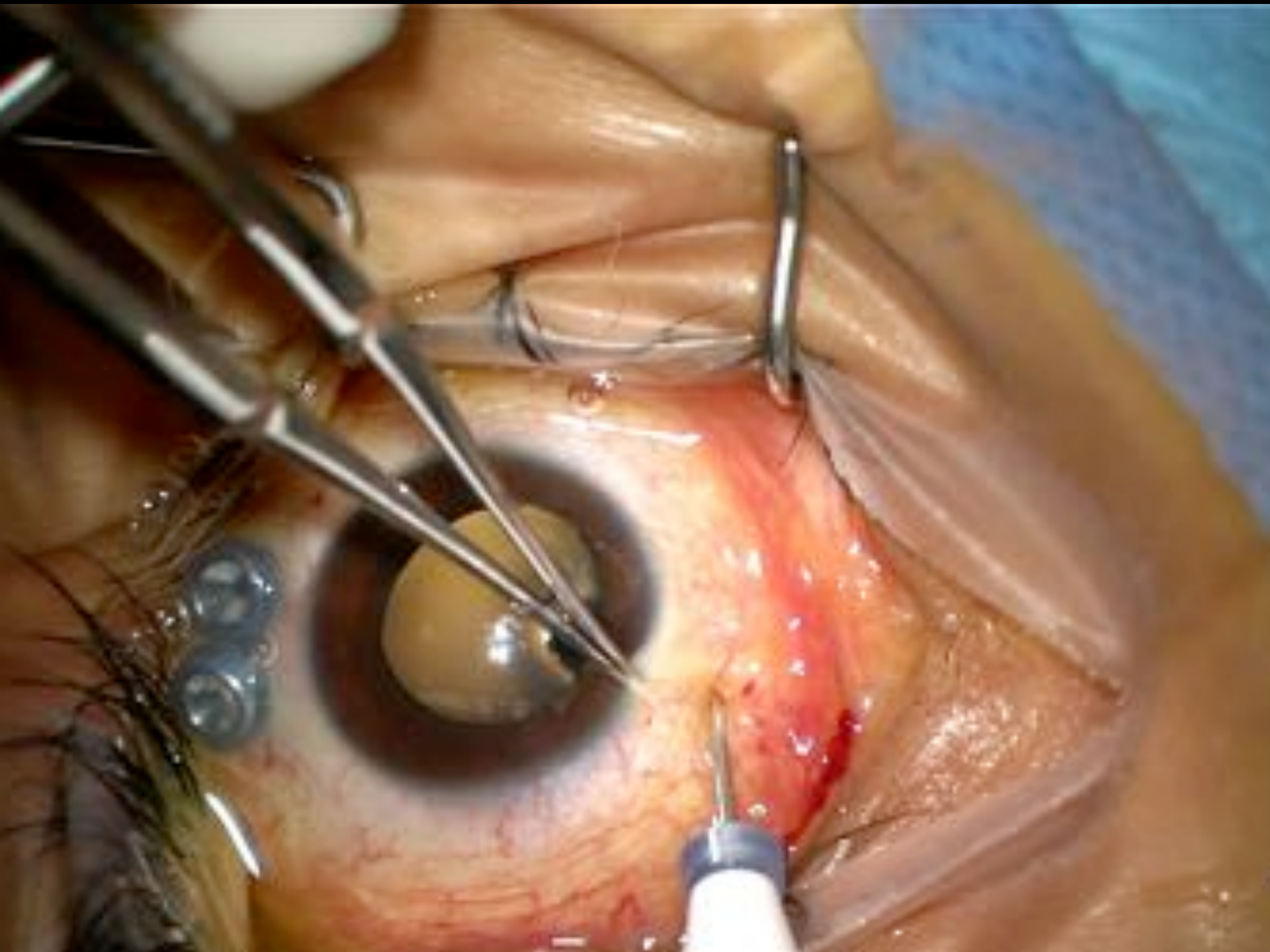
1 STK

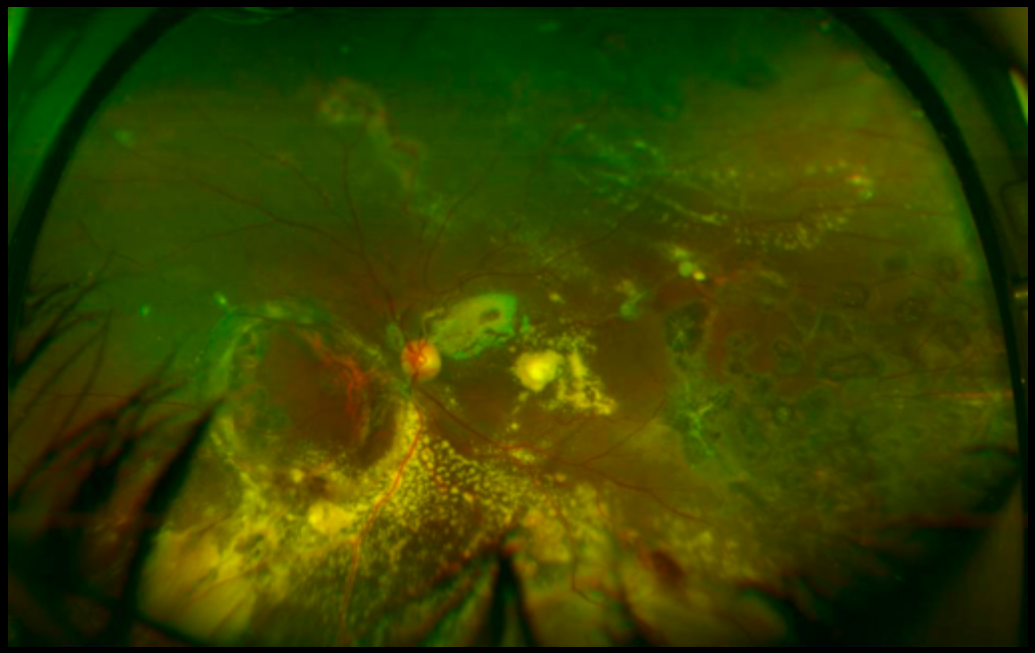


Yannuzzi NA, Tzu JH, Hess DJ, Berrocal AM. Retinoschisis in the setting of coats disease. Ophthalmic Surgery, Lasers and Imaging Retina. 2014 45(2); 172-174.

In globe salvage early PPV/EL/IVB/ STK controls disease



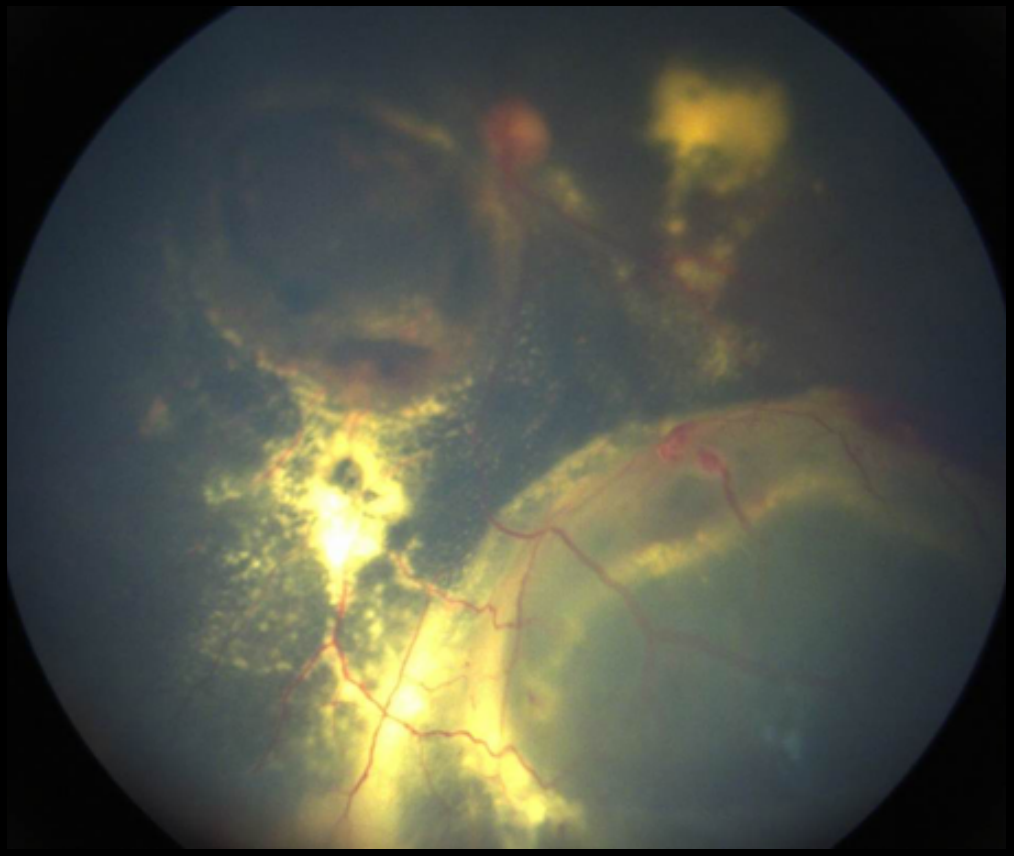




8 Months

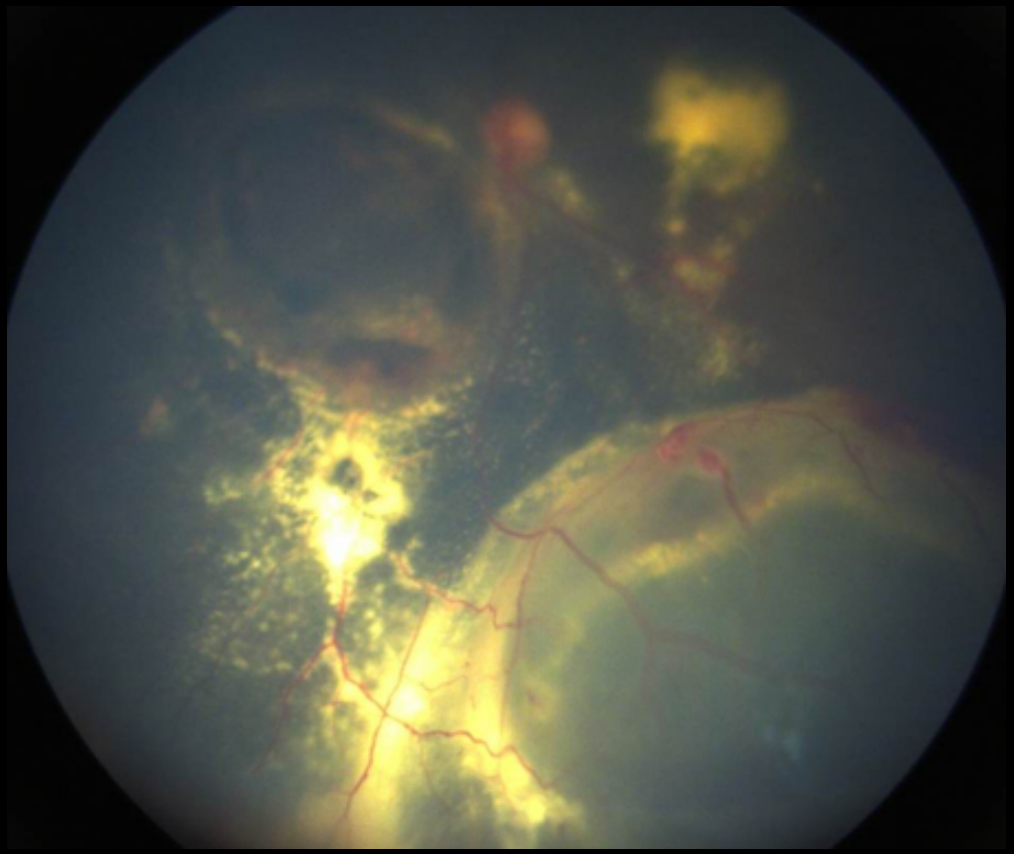
Methods

- Retrospective review with IRB approval
  - January 2014 to May 2018
- Identified:
  - 138 subjects with Coats
  - 133 had color images
- 18 subjects with retinoschisis (14%)

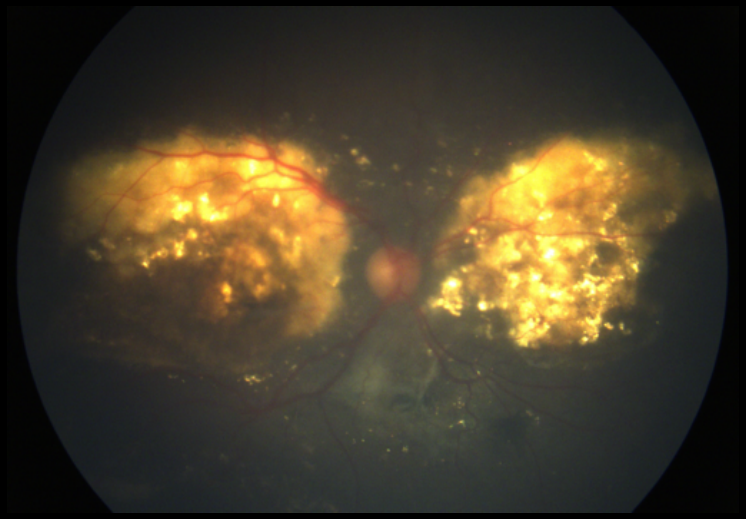


Methods

- Included:
  - Patients with more than 5 month follow up
  - Color images
- Two groups:
  - Schisis group
  - Control group



Results:



- Gender:
  - 75% male and 25% women
- Median age in the control group:
  - 17.8 years of age +/-15
- Median age in the schisis group:
  - 6.5 +/- 5

Results: Complete schisis control

	Schisis	Control
Diode Laser Photocoagulation Sessions	4.8± 2.9	n/a
Intravitreal Anti-VEGF	4.5 ± 2.9	n/a
Sub-Tenon's Injections Triamcinolone	1.70 ± 1.20	n/a
Intraocular Surgery (PPV or SB)	39% (P=0.14)	22 %

Discussion:

- Retinoschisis is a rare entity in Coats Disease
- The pathophysiology of retinoschisis in Coats Disease is not completely understood could be exudative or tractional (combined)
- Angiography guided laser and multimodal imaging are necessary to monitor progression of disease
- Early PPV may be indicated in the setting of rapidly progressive schisis

Thank you!

- Linda Cernichiaro-Espinosa
- Nathan Scott
- Timothy Murray
- William Harbour
- Sander Dubovy
- Catherine Negron