CME in a child

Carol Shields MD
Ocular Oncology Service
Wills Eye Hospital
Philadelphia PA USA
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
retina edema + exudation
Slight irregularity of the perifoveal capillaries with trace reduction in capillary vascular density
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
- Traction
- Dystrophy - Retinitis pigmentosa
- Degeneration

Check the periphery

Let’s look further ...
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
Two early reports on classification.
Coats disease classification

Stage 1  Telangectasia
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

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 1</td>
<td>T</td>
<td>1%</td>
</tr>
<tr>
<td>Stage 2</td>
<td>T E</td>
<td>14%</td>
</tr>
<tr>
<td>Stage 3</td>
<td>T E S</td>
<td>68%</td>
</tr>
<tr>
<td>Stage 4</td>
<td>T E S G</td>
<td>15%</td>
</tr>
<tr>
<td>Stage 5</td>
<td>T E S G P</td>
<td>2%</td>
</tr>
</tbody>
</table>

150 cases


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

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 (<3 vs. 3-10 vs. >10 years).

Results: The youngest age group had more referral diagnoses of retinoblastoma (29% vs. 15% vs. 0%, p < 0.01), worse presenting visual acuity (<20/200: 80% vs. 67% vs. 31%, p < 0.01), more advanced Coats disease stage (stage 3B: 65% vs. 38% vs. 10%, p < 0.01), and greater clock-hour extent of telangiectasia (7 vs. 5 vs. 4, p < 0.01), light bulb aneurysms (7 vs. 4 vs. 3, p < 0.01), exudation (10 vs. 7 vs. 5, p < 0.01), and vascular disorder characterized by retinal telangiectasia, micro and macro “light bulb” aneurysms, and intraretinal and subretinal exudation.

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.


Regarding Coats disease per age
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

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) and no light perception (n=5).4

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

- Stage 1
- Stage 2A
- Stage 2B
- Stage 3A1
- Stage 3A2
- Stage 3B
- Stage 4
- Stage 5

Visual outcomes:
- 20/20-20/40
- 20/50-20/200
- 20/400-NLP

Statistical significance: $p<0.001$
... back to our case
Summary

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