

COATS' DISEASE

INFORMATION SHEET

FOR PATIENTS
& FAMILIES

WHAT IS COATS' DISEASE?

Coats' Disease is a rare disorder characterized by abnormal development of the blood vessels in the retina. The retina is tissue lining the back of the eye that transmits light images to the brain and allows a person to see. In Coats' Disease, the blood-rich retinal capillaries leak fluid into the back of the eye. The leakage causes the retina to swell and may lead to partial or complete retinal detachment which can cause vision loss. Coats' Disease is almost always unilateral (impacting only one eye); in rare instances, both eyes may be affected but the symptoms are typically more severe in one eye. Coats' Disease was described in 1908 by Dr. George Coats. The specific cause of Coats' Disease is not known. It is not hereditary and no genetic marker has been identified yet. There is currently no cure. In the U.S., a rare disease is any disease, or disorder affecting fewer than 200,000 people in the United States.

Coats' Disease primarily afflicts children; two-thirds of patients are diagnosed before the age of 17. The average age at diagnosis is 8-16 years, although the disease has been diagnosed in patients as young as four months. Approximately one-third of patients are 30 years or older before symptoms begin. Coats' Disease is seen predominately in males (75%).

SYMPTOMS

Every patient is different; some may not exhibit any obvious signs and the patient may not always notice a change in vision in one eye if the other eye compensates for it. Experts agree that children who exhibit the following early warning signs should seek an evaluation immediately from an optometrist or ophthalmologist:

- **Eye turning outward or inward** (called strabismus).
- **Yellow or white reflection in flash photography** (called leukocoria). Just as the red-eye effect is caused by a reflection off blood vessels in the back of a normal eye, an eye affected by Coats' will reflect yellow or white in photographs as light reflects off cholesterol deposits.
- **Signs of loss of depth perception and parallax.** When an eye is affected by Coats', the unaffected eye will often compensate for the loss of vision, resulting in some loss of depth perception and parallax.
- **Deterioration of eyesight in either the central or peripheral vision.** This deterioration is likely to begin in the upper part of the vision field, as this corresponds with the bottom of the eye where blood usually pools.

Over time, Coats' Disease may cause detachment of the retina and substantial loss of vision. Additional signs may appear as Coats' Disease progresses, including elevated pressure inside the eye (glaucoma), clouding of the lens of the eye (cataract), reddish discoloration in the iris due to the growth of new blood vessels in the iris (rubeosis iridis or neovascular glaucoma), shrinking of the affected eyeball (phthisis bulbi), and/or inflammation of eye (uveitis).

PATH TO DIAGNOSIS

Children typically first see their pediatrician, who then refers the patient to an ophthalmologist. They will then refer the patient to a pediatric retina specialist if Coats' is suspected. Adult patients typically see an Ophthalmologist for an initial consultation and should be referred to a retina specialist.

To identify an Ophthalmologist with experience in treating Coats' Disease, visit www.coatsdiseasefoundation.org/coats-disease/find-a-doctor/.

Coats' Disease is divided into 4 stages, from the mildest to most advanced. Because Coats' often presents at a very advanced stage, it can be mistaken for Retinoblastoma, which is an extremely rare malignant tumor or form of cancer that develops in the retina. Retinoblastoma patients are usually diagnosed before the age of three. Common presenting symptoms include the white reflex in the pupil (leukocoria) and strabismus or eye misalignment. The presentation of symptoms in Retinoblastoma can be identical to those in Coats' Disease, therefore it is very important to correctly distinguish Coats' Disease from Retinoblastoma, since untreated Retinoblastoma can be life-threatening. Differentiating Coats' from Retinoblastoma often requires an examination under anesthesia and a computed tomography (CT) scan of the eyes and brain.

TREATMENT

The treatment of Coats' Disease is directed toward the specific signs present in each individual. A procedure that uses laser energy to heat and destroy abnormal blood vessels (photocoagulation) and/or a procedure that uses extreme cold to create a scar around the abnormal blood vessels (cryotherapy) are used singly or in combination to treat Coats' Disease. In conjunction with these procedures, steroids or other medicines such as bevacizumab may be injected into the eye to control inflammation and leaking from blood vessels. Surgery to reattach the retina may also be necessary.

If caught early, some level of vision can typically be restored. If caught in late stages, complete loss of vision can occur. Repair of retina previously damaged by the disease is not possible. In its final stages, enucleation (removal of the affected eye) is a potential outcome.



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QUESTIONS TO ASK YOUR DOCTOR

At the Jack McGovern Coats' Disease Foundation, we are often contacted by anxious parents or patients who are seeking information after receiving a diagnosis of Coats' Disease. The questions below are provided as a resource to assist you as you and your doctor decide the best approach for treatment. These questions do not constitute any form of medical advice or diagnosis. Each patient is unique. An experienced retinal specialist who has examined the patient is the best source of information for diagnosis and treatment. We always recommend getting a second opinion. To identify an Ophthalmologist with experience in treating Coats' Disease, visit our web page at www.coatsdiseasefoundation.org/coats-disease/find-a-doctor/.

BEFORE YOU BEGIN

Ahead of any treatments or procedures, it is important to ask yourself and the doctor the following questions:

- How do you know that this is Coats' Disease?
- Has your doctor treated other patients with Coats' Disease?
- Have you sought a second opinion? If not, please consult our Doctor Directory for knowledgeable doctors in your area. (see link above)

COATS' DISEASE QUESTIONS TO ASK YOUR DOCTOR

1. What Stage of Coats' Disease is he/she in?

- Will his/her vision get worse over time?
- Will the eye have pain?
- Will his/her eye start to turn out? Is muscle corrective surgery an option?
- Are cataracts likely?
- How likely is glaucoma? (due to retinal detachment)
- Is there calcification?
- What is the anticipated disease progression?

2. Is there a thorough vision exam available?

- Where is the vision affected? (central /peripheral/ distance)
- Does he /she have depth perception? (3D visibility)
- What about the non-Coats' eye?
- To what extent is his / her vision affected?
- Will we be able to use this as a baseline to measure progress/decline?

3. If there is a retinal detachment:

- What percent is the retina detached?
- Will it continue to detach?
- How quickly can it detach?
- Can you treat with laser or freezing treatments (also known as cryotherapy)?
- Is reattachment surgery advised?
- What are the risks associated with reattachment surgery? (i.e. further vision loss/re-detachment/success rate?)
- Will you be doing retinal mapping to compare progress with laser/cryotherapy?

4. Is laser/cryotherapy always successful?

- What are the risks with scarring?
- What about alternatives such as Avastin / Lucentis, which can help prevent new blood vessel growth?
- Is there a risk when using these anti-VEGF's to the rest of the body?

5. Is shrinking of the eye something to watch?

- What about pain in the eye – is that something to be anticipated? If so, would that indicate a need for removal of the eye (enucleation)?
- What is the difference between eye pain and an eye ache?

6. In his/her Non-Coats' eye, what will you be looking for?

If this isn't Coats' now, can we be confident it won't be in the future?

7. What are the cutting edge treatments on the horizon?

- Scleral shell to cover cataract or turning eye? Retinal Chip? Orbital eye implants? Stem cells to regenerate retinal tissue?

8. What is his / her IOP (Intraocular Pressure)?

- 12-18=normal, under 9 and over 20 a concern?
- What can affect his IOP?
- How can I tell if his IOP is changing?
- What drops are recommended at that point? (Cosopt/Atropine/Iopidine/Generic)?

9. Do you recommend glasses or protective eyewear to prevent injury to his/her non-Coats' eye?

- Polycarbonate Lens glasses? Tinted glasses?
- How often should he/she wear them?
- Should we restrict any sports or activities? Swimming after an EUA (exam under anesthesia)?

10. School recommendations?

- Educate staff and other kids/PE & recess?
- Where is the best place to sit in the classroom?

11. Are there vitamins specifically for the eye?

- What are the benefits of patching therapy and what the intent of patching? Vision therapy?

12. Should both eyes be dilated at each visit? Why?

- Dilating the pupils – how long does it last?
- Will the yellow glow in pictures go away with treatment?

13. What other doctors / specialists do you work with?

- Could his / her Coats' Disease be a symptom of another disorder? (FSHD, FEVR, Retinoblastoma)
- What resources do you have available for newly diagnosed patients/parents?

14. At what age do you stop doing EUA (exam under anesthesia)?

- What is the purpose of an EUA and what can be done during that time?