COATS' DISEASE
INFORMATION SHEET

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.

Jack McGovern
Coats' Disease Foundation

888.314.8853
contact@curecoats.org
www.coatsdiseasefoundation.org