



Fact Sheet

Coats' Disease

Information Retrieved from Medicine Met -

http://www.medicinenet.com/coats_disease/article.htm and Coat's Disease Foundation -

<http://www.coatsdiseasefoundation.org/>

What is Coats' Disease? Dr. George Coats in 1912 described a particular form of exudative retinitis (inflammation of the retina of the back of the eye), characterized clinically as follows: (a) occurrence in infant or young males; (b) involvement of one eye (unilateral); (c) absence of a known systemic disease; (d) exudates below the retinal vessels; (e) evidence of retinal hemorrhages; and (f) slow progression to retinal detachment, cataracts (cloudiness of the lens of the eye), optic nerve atrophy; and (g) glaucoma. In simpler terms, Coats' Disease involves atypical development in the blood vessels behind the retina of the eye. The blood-rich retinal capillaries of the retina break open and leak the serum portion of the blood into the back of the eye(s). The leakage causes retinal swelling and may lead to partial or complete detachment of the retina.

What are Symptoms and Signs of Coats' Disease? Early warning signs of Coat's Disease may include; (a) *yellow-eye in flash photography* - an eye affected by Coats' Disease will glow yellow in photographs, as light is reflected off cholesterol deposits; (b) a yellow reflex from the pupil (called leukocoria); (c) signs of lost depth perception; and/or (d) deterioration of vision in either the central or peripheral (side) vision. This deterioration of vision often begins in the upper part of the visual field, as this area of field corresponds with the bottom of the eye where blood usually collects. A final warning sign may be an eye turning out or in (called strabismus). There is a gradual decrease of vision, which may not be recognized at first due to the young age at which the disease begins. Coat's Disease is a painless disease unless there are other complications such as glaucoma (increased ocular pressure). Since Coats' Disease typically begins in one eye, the child may compensate well and no one may observe that vision is impaired in the affected eye. The vision loss may begin as early as infancy, but is most commonly seen between 6-9 years of age.

How is Staging of Coats' Disease Classified? There are five stages to Coat's Disease. Stage 1 is characterized by telangiectasia only, which involves chronic dilation of groups of capillaries causing elevated dark red blotches on the skin. Stage 2 results in telangiectasias and exudation

and is further subcategorized depending on involvement of the fovea (center of the retina with the sharpest vision). Stage 3 results in subtotal retinal detachment, also subcategorized based on foveal involvement. Stage 4 involves a total retinal detachment with the presence of glaucoma. Stage 5 is end-stage disease with a blind, painless or painful eye, and total retinal detachment, often with cataract and eventual phthisis bulbi (shrinkage of the eyeball after trauma or disease).

What are Causes and Risk Factors for Coats' Disease? There are no known causes or risk factors of Coat's Disease. There is no known hereditary component or any other cause. Although there may be some evidence that Coats' Disease is caused by a somatic mutation of the Norrie disease protein (NDP) gene. Coats' Disease occurs in males three times as often as in females. Coat's Disease does not appear to be inherited and has no known racial or ethnic association. It cannot be prevented.

What is the Prevalence of Coat's Disease? This is a rare disorder. Fewer than 200,000 people in the population of the United States are affected by Coat's Disease. One study found 0.09 cases in every 100,000 individuals. It has been estimated that about 69% of the cases are male. The average age at diagnosis is reported to be 8–16 years, although this disease has been diagnosed in infants as young as four months. The peak age of onset is between 6-8 years of age, but can range from four months up to 71 years. About two-thirds of childhood cases show symptoms before age 10 years; approximately one-third of individuals with Coats' Disease are 30 years or older before the symptoms begin.

How is Coats' Disease Diagnosed? Although Coat's Disease may be diagnosed on a routine eye exam, it often is not suspected until a later stage when a child develops a cloudy white or yellow pupil due to the presence of a cataract or a retinal detachment. It can be observed in a photograph where the pupil of the affected eye appears yellow or white, while the other pupil appears to be normal black. Coats' Disease may be diagnosed on a routine eye examination, when the pupils are dilated, and a cataract is seen or abnormal blood vessels may be noted at the back of the eye. Retinal damage may be observed due to the accumulation of cholesterol deposits in the vessels. The eye care specialist will also determine whether there is coexisting optic nerve atrophy or glaucoma.

Since Coats' Disease may come on gradually in one eye, the person often doesn't notice the painless decrease of vision. As it progresses, the person may notice the following symptoms from the retinal damage: loss of peripheral (or side) vision, light flashes, floaters (deposits or condensation in the vitreous jelly of the eye), or a general loss of vision. A parent of a young child may notice a yellow reflex in the pupil (due to a cataract or retinal detachment) when a picture is taken.

Coats' Disease may be misdiagnosed, so it is important for parents to work with an eye care specialist who is experienced in managing the disease. One challenge to making an accurate diagnosis is that the patients are often too young to describe their physical problem or may not understand the change in their vision. Also the presenting symptoms of Coat's Disease can mimic other conditions such as retinoblastoma (cancer of the brain/eye), which is potentially life-threatening. Typically individuals with Coats' Disease do not experience any other health problems. Though it is important to note, symptoms typical of Coat's Disease may also appear

in several multisystem disorders, such as fascioscapulothoracic muscular dystrophy, pericentric inversion of chromosome 3, and Alport syndrome.

What is the Treatment for Coats' Disease? This is a progressive disease with no obvious visual impairment in the early stages of the disease. If caught in an early stage (relatively rare since it usually isn't recognized by the child), freezing or laser treatment of the retina may be successful. Occasionally the disease stops on its own and may even reverse itself. The medical treatment is designed to halt the blood vessel progression. Once Coat's Disease is diagnosed, an eye care specialist should follow the person on a regular basis. It is important that the patient have regular examinations of the good eye to ensure its maintained health.

What are the Educational Implications? Children with Coat's Disease may be eligible for early childhood or school-age special education services, if the associated vision loss prevents the child from receiving reasonable educational benefit from general education. The child may benefit from specially designed instruction and equipment. The child's hearing should be screened to ensure that there are no further sensory complications to learning.

Where to Get More Information about Coats' Disease

The Jack McGovern Coats Disease Foundation <http://www.coatsdiseasefoundation.org>

The Coats' Disease web site has a wealth of information about the disease for families:
<http://www.coatsdisease.org/index.html>

For more information about the CO Services for Children and Youth with Combined Vision and Hearing Loss Project contact:

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Fact Sheets from the Colorado Services to Children and Youth with Combined Vision and Hearing Loss Project are to be used by both families and professionals serving individuals with vision and hearing loss. The information applies to children, birth through 21 years of age. The purpose of the Fact Sheet is to give general information on a specific topic. The contents of this Fact Sheet were developed under a grant from the United States Department of Education (US DOE), #H326C080044. However, these contents do not necessarily represent the policy of the US DOE and you should not assume endorsement by the Federal Government. More specific information for an individual student can be provided through personalized technical assistance available from the project. For more information call, (303) 866-6681 or (303) 866-6605. Updated: 9/12