

# Coats' Disease

## WHAT IS COATS' DISEASE?

Coats' disease is caused by a problem with the arteries and veins (blood vessels) inside the eye that provide blood and oxygen to the retina. These abnormal blood vessels are usually located in the temporal retina. In Coats' disease, the blood vessels are dilated, abnormally twisted and leaky. This prevents the normal flow of blood and allows fluid to leak out of the blood vessels causing a buildup of fatty (lipid) material into the retina. If a large amount of fluid (exudate) builds up, it can cause a detachment of the retina and loss of vision. [See figures 1 and 2]. This condition typically affects the vision in only one eye, however newer tools have shown a number of patients have silent involvement of the second eye. It is most commonly seen in males. There is no genetic or inherited basis for this condition.



**Fig. 1:** Retinal detachment visible through pupil in patient with Coats' disease.

## WHAT ARE SIGNS AND SYMPTOMS OF COATS' DISEASE?

Presenting symptoms in children could be leukocoria, which is an abnormal white light reflection from the pupil of the eye, decreased vision, an eye that crosses inward or turns out, and pain from increased eye pressure in severe cases. Clinical signs include abnormal blood vessels seen in the peripheral retina, retinal exudates/edema, and retinal detachment.



**Fig. 2:** Retinal photograph showing hemorrhages and exudates from leaking blood.

## HOW IS COATS' DISEASE DIAGNOSED?

Coats' disease is diagnosed by direct visualization of the retinal vessels, and special imaging modalities such as ultrasound, fluorescein angiography, optical coherence tomography (OCT) and OCT angiography (OCTA).

## WHAT ARE THE STAGES OF COATS' DISEASE?

The disease progress is divided into five stages. In stage 1, abnormal blood vessels are seen in the retina but these vessels are not yet leaky. Stage 2 of the disease is characterized by leakage from the vessels into the retina. The effect on vision depends on the amount of fluid that has leaked into the retina, the size and location of the area involved. If only a small amount of fluid has leaked, and the center of the retina is not involved, the vision may remain normal. If large amounts of fluid leak and/or the center of the retina is involved, vision loss can be severe. The accumulation of fluid leaking from the vessels can cause the retina to detach. The presence of such detachment takes the disease to stage 3. If this is further complicated with glaucoma (raised pressure of the eye), the disease classifies as stage 4. Stage 5 is the end stage of the disease when the eye becomes blind and may be painful due to the raised pressure inside the eye.

## HOW IS COATS' DISEASE TREATED?

Treatment is sometimes recommended to prevent progression of the disease. Better results are achieved when the disease is treated in the earlier stages. Laser or freezing treatments (cryotherapy) are often utilized to constrict the abnormal blood vessels and stop the leakage of fluid in Stage 2 or 3A. Anti-

vascular endothelial growth factor therapy, or anti-VEGF therapy, a newer medication given by direct injection into the eye, – can reduce fluid leak and retinal detachment. Vitrectomy surgery may be needed in the advanced stages of the disease to treat more extensive retinal detachments. Continued monitoring is necessary to check for recurrence of the disease after treatment, and for involvement of the second eye. Glasses and patching therapy for amblyopia are sometimes necessary. The prognosis of this condition depends on various factors, including the patient’s age and stage of disease at diagnosis, and the rate of disease progression. In patients presenting younger than three years of age, the disease is usually more aggressive. The majority of patients often have poor vision in the affected eye. However when patients present at an older age, the disease is often milder.

Rao, Prethy MD, MPH<sup>†</sup>; Knapp, Austen N. BS<sup>†</sup>; Todorich, Bozho MD, PhD<sup>†</sup>; Drenser, Kimberly A. MD, PhD<sup>†</sup>; Trese, Michael T. MD<sup>†</sup>; Capone, Antonio Jr MD<sup>†</sup> Anatomical Surgical Outcomes of Patients With Advanced Coats Disease and Coats-Like Detachments: Review of Literature, Novel Surgical Technique, and Subset Analysis in Patients With Facioscapulohumeral Muscular Dystrophy, *Retina*: October 2019 - Volume 39 - Issue - p S182-S190 doi: 10.1097/IAE.0000000000002463

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