



## Coats' Disease, The 2018 Brief

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**Received date:** 19 March 2019; **Accepted date:** 30 March 2019; **Published date:** 05 April 2019

**Citation:** Selim E, Selim M, RehabAuf, "Coats' Disease, The 2018 Brief". *Asp Biomed Clin Case Rep*, vol.2, no.1: 10-12, 2019.

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### Abstract

**Purpose/Aim of the study:** Authors planned to study articles published in 2018 to find out the latest update on this rare condition.

**Materials and Methods:** Pubmed search was conducted with 2018 in focus as a year of publication including only articles on humans and in English language.

**Results:** out of 22 articles published in 2018, only 7 were found relevant on human subjects and in English language.

**Conclusions:** Newer treatment modalities were tried in 2018 as well as the old well approved modalities.

### Keywords:

Coats; Exudative; Retinal; Detachment; Photodynamic

### Introduction

Coats disease is a condition characterized by dilated retinal vessels with intra and subretinal exudation [1]. Coats [2] first described the condition as one of the forms of retinal diseases with massive exudation in 1908. The condition is known to be idiopathic affecting one eye of male patients. Various other ophthalmologists described the condition originally observed by the Scottish ophthalmologist, Coats, and later modified his observation of the disease. Leber [3] claimed he observed retinal aneurysms that did not share the massive exudation observed by Coats. However, he later admitted they were early stages of Coats disease prior to exudation that was the original feature Coats described with various degrees of vascular dilatation. Von Hippel managed to separate

the most advanced form of exudation Coats described to be a distinct entity, later called **retinal hemangioblastoma**. Nowadays, the term "**Coats disease**" is used to refer to a large array of idiopathic unilateral retinal telangiectasia affecting males [4]. Among those is Foveal telangiectasia type 1 which was first described by Gass and Boldi [5] in 1993. The condition as they described it was a category of idiopathic juxta foveal telangiectasia (IJFT) originally described in 1982 but occurred more in males as a congenital condition.

### Methods

We used PubMed as the main resource to search medical literature. Search was conducted to identify all articles published on Coats disease in the year

2018. Keywords used were Coats disease. It is the author's knowledge that Coats disease is a rare disease and hence not many articles were expected for the period of one year. Hence, the broad term Coats disease was used to include every single publication that mentioned the term in the past calendar year. Only articles with English language abstracts were included. We intended to exclude any animal based study. No attempts were made to include any unpublished data.

## Results

The search revealed 22 records. After applying exclusion criteria, 12 abstracts were screened. Of those screened, 7 articles were found to be relevant. Out of those, 4 articles discussed treatment of Coats disease either with surgery, laser or injections. While four articles discussed appearance, clinical findings and other investigative modalities proved helpful in Coats disease.

## Discussion

Although Coats disease has always been believed to be a unilateral disease, Jung and colleagues [6] were able to demonstrate some vascular abnormalities by Fluorescein Fundus angiography in the fellow eyes when studying a series of 47 patients with Coats disease.

A case was described in a 30-year-old female with peripheral vascular sheathing. However, with Fluorescein Fundus Angiography multiple microaneurysms were seen which appeared like Leber military aneurysms. The lady's macular edema did not respond to Anti-vascular endothelial growth factors (Anti-VEGF) and responded to Triamcinolone injection indicating an inflammatory process [7]. Another case report by Fernandes et al [8] described Coats in a young girl. Those two cases are a proof that Coats disease which affects predominantly males can also affect females. Moreover, it shows that an inflammatory element exists in Leber's military aneurysm as shown by the presence of sheathing as well as the positive response to triamcinolone injection. Another case was reported where vasculitis was observed in a 22-year young man who present with counting finger vision with exudative

detachment and multiple microaneurysms [9]. His condition stabilized with Anti-VEGF compare to lack of response to Anti-VEGF in the previous case reported by Lee et al [7].

A retrospective study by Zhang and colleagues [10] on 28 patients' medical records showed that Anti-VEGF offered visual improvement when used either alone or in combination with laser therapy. This method of treatment was a safe one that did not have any significant side effect.

Photodynamic therapy was proved to be efficient in treating Coats disease when leaking blood vessels are close to the posterior pole. This treatment modality was able to cure an eye of a 15 years old boy with an initial presentation of exudative retinal detachment and a visual acuity of 20/100 which improved to 20/20 in the 10 months follow up following treatment with photodynamic therapy which is safer to use at the posterior pole than the traditional treatment of Coats disease such as cryotherapy and laser photocoagulation [11].

Kusaka [12] reports treating patients with severe exudative retinopathy by external drainage whether with or without vitrectomy. This drainage is normally followed by the traditional cryotherapy of laser photocoagulation. It is believed to be associated with better outcome than applying cryotherapy to parts of the retina that are highly elevated by exudation which are also less likely to respond to Anti-VEGF.

## Conclusion

Although Coats disease has always been believed to affect one eye in males predominantly, recent studies in 2018 reported the disease in females and features of the disease in the other eye. The traditional wisdom for treating Coats disease has always been believed to be photocoagulation and/or cryotherapy depending on the location of leaking blood vessels. However, with the presence of the disease more centrally and with the inclusion on Macular Telangiectasia type 1 as a variety of the disease, less aggressive mode of treatment were needed. Anti-VEGF provided a safe and effective way to treat Coats disease. Photodynamic Therapy was used and found to be

effective. Another challenge was addressed which was eyes with severe exudation in which laser treatment or cryotherapy is not likely to work because the retina is highly elevated by fluid. Drainage of that fluid externally with or without vitrectomy addressed this challenge.

### Conflict of interest

Authors declare that there is no conflict of interest.

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