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# Intraoperative Fluorescein Angiography–Guided Treatment in Children with Early Coats' Disease

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***Purpose:*** To review the anatomic and visual outcomes of a series of children diagnosed with Coats' disease and treated on the basis of intraoperative fluorescein angiography (FA) findings.

***Design:*** Retrospective case series.

***Participants:*** Twenty-five children 2 to 15 years of age diagnosed with early Coats' disease and treated after intraoperative FA.

***Methods:*** Retrospective review of patients who underwent treatment for stage 2 Coats' disease in a tertiary center in the United Kingdom between 2007 and 2012. The children underwent treatment to the telangiectatic vessels and associated areas of retinal nonperfusion identified on intraoperative FA performed with a wide-angle retinal camera (RetCam II).

***Main Outcome Measures:*** Anatomic and structural assessment of the retina after treatment, visual acuity (VA), and complications related to treatment.

***Results:*** Twenty children with Coats' disease who underwent intraoperative RetCam FA and retinal ablative treatment and who had more than 3 months of follow-up were identified from clinical records. All had unilateral disease. Six eyes were classified as stage 2a, and 14 eyes were classified as stage 2b. The median duration of follow-up was 21 months (mean, 26 months). Fifteen eyes needed 1 treatment to stabilize the disease, and 5 eyes needed a second treatment. None of the patients demonstrated progression of the disease to a more severe stage. Twelve eyes had a final VA of 0.4 logarithm of the minimum angle of resolution (logMAR) or better, 6 patients had VA between 0.4 and 1.0 logMAR, and 2 patients had VA worse than 1.0 logMAR.

***Conclusions:*** The treatment of retinal telangiectasia and areas of retinal nonperfusion identified by wide-angle intraoperative FA in children with stage 2 Coats' disease led to good anatomic outcome, with preservation of VA in most cases. *Ophthalmology 2015;122:1195-1202 © 2015 by the American Academy of Ophthalmology.*

Coats' disease is an idiopathic, unilateral disease characterized by telangiectatic vessels usually in the retinal temporal periphery with associated retinal exudation that may involve the macular region.[1,2] It was first described at the beginning of the 20th century by George Coats, who noted that males were affected more commonly than females.[3] Coats' disease is rare, with a reported incidence of 0.09 per 100 000 of the population.[4] It usually manifests in childhood with leukocoria, strabismus, or reduced acuity often detected during vision screening. It also may be found as an incidental finding during a routine ocular examination.[4]

Untreated Coats' disease may cause vision loss resulting from macular involvement, and in advanced disease, there may be exudative retinal detachment with secondary neovascular glaucoma.[5,6] Shields et al[7] classified Coats' disease into different stages (Table 1). This clinical staging is useful in predicting the visual outcome after treatment.

The primary goal of treatment is to stop the exudation from the abnormal vessels to prevent retinal detachment and neovascular glaucoma and, where possible, to maintain or improve visual acuity (VA). The standard treatment for Coats' disease is laser photocoagulation or cryotherapy to the telangiectatic vessels.[7,8] Where the retina is attached and the peripheral retinal exudation is not extensive, laser is the treatment of choice. Cryotherapy commonly is preferred when the exudation is extensive or the retina detached,[9,10] but some authors report promising results from laser treatment alone.[8,11] Using such treatment, most cases are reported to need more than 1 treatment to eradicate retinal exudation.[7,8,11] The approach to treatment has been different in adults and children. In adults with Coats' disease, retinal angiography is performed routinely and treatment is targeted specifically to the regions of abnormal retinal perfusion. In the pediatric population, treatment historically has been based on direct visualization of the telangiectatic vessels during examination under anesthesia. Recently, a portable wide-angle angiography system, the RetCam (Clarity Medical Systems, Pleasenton, CA), has become available for the assessment of retinovascular diseases in children.[12,13] It has proved useful in the investigation of retinoblastoma, retinopathy of prematurity, and other retinovascular abnormalities.[14,15] Although intraoperative wide-field fluorescein angiography (FA) is used by many centers treating pediatric retinal disease, there are only a few



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Table 1. Classification of Coats' Disease as Described in 2001 by Shields et al[7]

| Stage | Description |
|---|---|
| 1 | Retinal telangiectasia only |
| 2 | Telangiectasia and exudation |
| 2a | Extrafoveal exudation |
| 2b | Foveal exudation |
| 3 | Exudative retinal detachment |
| 3a | Subtotal detachment |
| 3a1 | Extrafoveal subtotal detachment |
| 3a2 | Foveal subtotal detachment |
| 3b | Total retinal detachment |
| 4 | Total retinal detachment and glaucoma |
| 5 | Advanced end-stage disease |

published reports of its use in Coats' disease.[8,16,17] We present our experience using RetCam FA instead of retinal examination alone (historical treatment) to guide treatment in a large series of children with early Coats' disease.

## Methods

### Patients

We identified 25 children with a diagnosis of Coats' disease who attended the pediatric retinal service at Moorfields Eye Hospital from December 2007 through March 2012 and underwent treatment for stage 2 Coats' disease after intraoperative FA using the RetCam II. This study has been approved by the Hospital Review Board and was performed in accordance with the Declaration of Helsinki. To be included in the study, the patients had to show idiopathic telangiectasia and lipid exudation in the sensory retina or the subretinal space in the retina periphery (outside zone 1 in retinopathy of prematurity staging classification[18]) with or without posterior pole involvement. Retinal telangiectasia was defined as irregular and dilated small or medium vessels.[2]

### Management

All the patients were first seen in the clinic, where color fundus photography and time-domain or spectral-domain optical coherence tomography (OCT) were attempted if the child was old enough to cooperate. Color fundus photography was performed with a TRC-50IA Retinal Fundus Camera (Topcon, Tokyo, Japan), time-domain OCT was performed with the 3D OCT 2000 (Topcon), and spectral-domain OCT was undertaken with the Spectralis with viewing module version 5.1.2.0 (Heidelberg Engineering, Heidelberg, Germany). The spectral-domain OCT protocol included a horizontal, linear scan (100 B-scans averaged to improve the signal-to-noise ratio) centered on the fovea where possible and a volume scan (minimum of 19 B-scan slices, 20×20°). If the clinical suspicion of Coats' disease was confirmed, an examination under anesthesia was arranged. The examination under anesthesia consisted of indirect ophthalmoscopy evaluation of the retina in both eyes and color photography and FA performed with the RetCam II. Each patient was given an intravenous bolus of 20% sodium fluorescein (0.1 ml/kg) before performing FA. The images initially were obtained from the affected eye before imaging the fellow eye.

On the basis of the fluorescein findings, indirect laser was administered to the areas of retinal nonperfusion and the telangiectatic vessels were treated directly with laser or cryotherapy. Cryotherapy was used only when there was confluent extensive exudation in the area of vascular abnormalities. The treatment was administered to all patients by the same clinician (A.T.M.). For all patients treated with laser, a diode-pumped, frequency-doubled yttrium–aluminum–garnet Laser Litechnica LightLas 532 laser (wavelength, 532 nm; Litechnica Ltd, Heston, UK) was used. After treatment, further color pictures with the RetCam were obtained.

The patients first were reviewed between 4 and 12 weeks after the treatment in clinic, when color photography and OCT were performed, if possible. Subsequently, the patients were monitored in the clinic every 3 to 4 months. Further treatment was undertaken if there was a lack of resolution or an increase of exudation at follow-up appointments. Patients younger than 7 years underwent occlusion therapy of the unaffected eye to treat any element of amblyopia unless there was extensive macular damage. For the purposes of the study, we recorded demographic data, VA, and clinical findings (color fundus photographs and FA images) before and after surgery.

## Results

Twenty-five patients who had undergone intraoperative FA before treatment of Coats' disease were identified. Twenty had at least 3 months of follow-up after treatment and were included in the study. Table 2 shows demographic characteristics, age at presentation, and first clinical symptoms.

### Ocular Findings

Of 20 patients, 6 were classified as having stage 2a disease and 14 were classified as having 2b disease.[7] In stage 2a disease, all patients but 1 showed on clinical examination telangiectasia involving less than 60° of the peripheral retina. The exudation was posterior to the telangiectasia and more widespread. In all cases, FA demonstrated more widespread retinovascular changes, such as telangiectasia and microaneurysms, as well as capillary nonperfusion (Table 3).

In stage 2b disease, during fundus examination performed under anaesthesia, 13 patients had telangiectatic vessels over 90° or less, and 1 patient had 180° of telangiectasia. The exudation was present for 360° in 8 patients, for 180° to 210° in 2 patients, for 90° in 2 patients, and only at the posterior pole in 2 children (Table 3). Clinical examination of the fellow eye showed normal results in all cases.

Fluorescein angiography showed more extensive areas of telangiectasia and the presence of leaking microaneurysms in all but 1 case. All children showed changes in the capillary bed anatomic characteristics, such as coarsening or capillary bed closure. In 6 patients, the capillary abnormalities were present for less than 180°, and in 8 patients, they were present for 180° or more. Imaging of the fellow eye showed an area of far peripheral retinal nonperfusion of 4 clock hours in 1 subject. Examination of the retina was normal in each of his parents.

### Treatment

In all cases but one, it was possible to perform the treatment immediately after FA. One case needed to be rescheduled for treatment because extensive leakage of fluorescein into the vitreous made laser treatment difficult. Eighteen patients initially were treated with laser, and 2 patients were treated initially with cryotherapy because of confluent extensive exudation. Of those receiving laser treatment, the mean number of laser burns was 700 (range, 300–2000). Five patients received a second treatment because of evidence of residual telangiectatic vessels with persistent exudation; 4 patients were treated with laser and 1 patient was treated with cryotherapy.