*Original Manuscript*

ASRS American Society of Retina Specialists

Journal of VitreoRetinal Diseases
2025, Vol. 9(5) 659–666

DOI: 10.1177/24741264251355635
journals.sagepub.com/home/jvrd
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# Comparison of Adult and Pediatric Eyes With Coats Disease Using Multimodal Imaging

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## Abstract

**Purpose:** To compare the clinical features, imaging findings, and treatment outcomes between adults and children with Coats disease. **Methods:** This retrospective, multicenter, observational case series comprised adult and pediatric patients with Coats disease treated between 2016 and 2022. **Results:** The study included 19 eyes from 10 children and 8 adults. The median age at presentation in children was 6 years (range, 2-17) and in adults was 29 years (range, 24-61). Children presented at later stages of disease compared with adults. The median total clock hours of exudation were greater (2.5 vs 1, $P = .01$) in children, and macular optical coherence tomography (OCT) findings, including subretinal and intraretinal fluid, were more common (60% vs 11%, $P = .03$) compared with adults. Children were more likely to present with vessels traversing the foveal avascular zone on OCT angiography (OCTA) (75% vs 29%). At presentation and final follow-up, the visual acuity (VA) was lower in children compared with adults ($P \leq .01$). **Conclusions:** Upon diagnosis with Coats disease, children present at later stages and are more likely to have macular involvement and lower VA compared with their adult counterparts. OCTA showed more qualitative abnormalities in children, while the affected eyes of adults had a lower vessel density of the superficial and deep capillary plexuses compared with their fellow eyes.

## Keywords

Coats disease, optical coherence tomography angiography, pediatric

## Introduction

Coats disease is an idiopathic, nonhereditary retinal vasculopathy characterized by telangiectasias as well as intraretinal and subretinal exudates.[1] The risk for sight-threatening complications, including retinal neovascularization, exudative retinal detachment (RD), phthisis bulbi, and neovascular glaucoma, is greater in advanced stages of the disease.[2,3] Although typically presenting unilaterally in young men,[3–5] Coats disease has been described with bilateral involvement and can present in adulthood and in women.[3,6] Some reports have suggested that Coats disease presenting in childhood is associated with more severe disease manifestation and a worse visual prognosis.[5,7,8]

Although fluorescein angiography (FA) is considered the gold standard in the diagnostic workup for Coats disease, optical coherence tomography angiography (OCTA) is an emerging modality to evaluate retinal vascular pathology. OCTA allows for noninvasive detection of flow to visualize abnormalities of the vascular structure, particularly in the superficial and deep retinal vascular plexi.[9] Limited small studies have described OCTA findings in pediatric patients with Coats disease,[10–15] but there are no previous quantitative OCTA studies in adult patients. In addition, there is a lack of studies comparing OCTA imaging findings of the adult and pediatric populations. The aim of the current study was to evaluate the clinical features and outcomes between adult and pediatric populations with Coats disease, focusing on OCTA findings.

## Methods

### *Study Cohort*

This was a retrospective, comparative series comprising 10 children and 8 adults with Coats disease treated at 2 tertiary centers, New England Eye Center and Massachusetts Eye and Ear in Boston, MA. The study conformed to the tenets of the Declaration

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of Helsinki and was prospectively approved by the institutional review board of Massachusetts General Brigham (protocol ID# 2021P002833). The requirement for informed consent was waived due to the nature of the study. Subjects diagnosed with Coats disease who were treated between January 2016 and June 2022 were included. Patients with initial diagnosis and treatment at an outside facility as well as incomplete medical records or any history of other chorioretinal diseases were excluded. Patients were divided into 2 subgroups according to their age at presentation, a pediatric subgroup (disease diagnosis before 18 years of age) and an adult subgroup (disease diagnosis at age 18 years and older).

Data collected included demographics, best-corrected visual acuity (BCVA) in logMAR at presentation,[16] intraocular pressure, stage and disease laterality, symptoms at presentation, and imaging findings, including fundus photography, OCT, FA, and OCTA. Treatment modalities and outcomes, including retreatment and complications, were recorded.

Coats disease was staged according to the Shields classification into stage 1 (retinal telangiectasia only), stage 2a (telangiectasia and extrafoveal exudation), stage 2b (telangiectasia and foveal exudation), stage 3a (subtotal exudative RD), stage 3b (total exudative RD), stage 4 (total detachment and secondary glaucoma), and stage 5 (advanced end-stage disease).[17] The stage of disease was evaluated by an experienced ophthalmologist during the clinic visit, retrieved from the medical records, and subsequently confirmed through a review of the retinal imaging. Fundus photographs were reviewed for the presence or absence of telangiectasia, exudation, RD, subretinal fibrosis, vitreoretinal fibrosis, and vitreous hemorrhage. The extent of peripheral telangiectasia and intraretinal or subretinal exudation at presentation was quantified in terms of clock hours from 1 to 12. Macular OCT images were assessed for the presence of cystoid macular edema, subretinal fluid (SRF), and intraretinal fluid (IRF). OCTA images were obtained using the RTVue-XR Avanti (Optovue Inc), Zeiss Cirrus 5000-HD-OCT Angioplex (Carl Zeiss Meditec Inc), or the Plex Elite 9000 (Carl Zeiss Meditec Inc).

Primary outcomes included clinical features such as change in the VA between presentation and final visit and imaging findings comparing the adult and pediatric subgroups. Secondary outcomes included treatment regimens with retreatment rate and complications.

### *Imaging Protocol and Analysis*

The highest signal strength 3 × 3 mm macular OCTA scan of both the superficial capillary plexus and the deep capillary plexus was analyzed for affected and fellow eyes. After projection removal, en face retinal slabs were exported using the built-in review software of the OCTA devices. Images featuring motion artifacts or gap artifacts, which would hinder the image processing, were excluded.

Fiji software (NIH) was used for image processing. Calculations of the vessel density of the superficial and deep capillary plexi were performed after image binarization using the default method.[18] Furthermore, each binarized 3 × 3 mm OCTA image was divided into 4 quadrants, centered on the fovea: the superotemporal quadrant, the superonasal quadrant, the inferotemporal quadrant, and the inferonasal quadrant. The contour of the foveal avascular zone (FAZ) was manually outlined with the ImageJ Freehand Selection Tool and the FAZ area calculated. All measurements were performed independently by 2 observers (S.H., A.Y.) and the average values reported.

### *Statistical Analysis*

A descriptive analysis of outcomes was performed and is presented as the mean ± SD or median with interquartile range (IQR) for continuous variables and percentages for categorical variables. All variables were tested for normality using the Shapiro–Wilk test. A comparison of continuous variables was performed using the Mann–Whitney nonparametric test, while categorical data were analyzed using a $\chi^2$ test. All statistical significance levels were set at $P < .05$. Analysis and descriptive statistics were performed using SPSS (version 28, SPSS Inc).

## Results

### *Demographic Characteristics*

Nineteen eyes from 18 patients were included in the study, with 10 patients (10 eyes [53%]) in the pediatric subgroup and 8 patients (9 eyes [47%]) in the adult subgroup. Patient characteristics are summarized in Table 1. The median age at presentation was 6 years (IQR, 4-8) in the pediatric subgroup and 29 years (IQR, 27-43) in the adult subgroup. Although 50% of the pediatric patients (median age, 8 years; range, 5-17) presented with decreased vision, 30% of the children (median age, 4 years; range, 2-7) were asymptomatic; 56% of adults were asymptomatic at diagnosis, and 44% of adult patients presented with decreased vision. Asymptomatic cases were referred from ophthalmologists or optometrists after incidental findings during routine eye examinations. The median initial VA was lower in children (logMAR BCVA 1, IQR, 0.54-1.40, or Snellen 20/200) than adults (logMAR BCVA 0, IQR, 0-0.32, or Snellen 20/20) ($P < .01$).

### *Clinical Characteristics*

Although no pediatric patient presented with stage 1 disease, adult patients were diagnosed at an early stage (stage 1, 2a, and 2b), and none had stage 3a or 3b disease (Table 1). The median total clock hours of exudation were greater in children (2.5, IQR, 2-12) compared with adults (1, IQR, 0-2) ($P = .01$). The macular OCT at presentation was abnormal in 60% of children (40% with SRF and 20% with IRF) compared with 11% of adult patients (only 1 adult had macular edema, SRF, and IRF) ($P = .03$). The FA findings were similar between the groups (Table 2). Figure 1 shows representative fundus photographs

**Table 1.** Comparison of Demographic and Disease Characteristics at Presentation Between Children and Adults With Coats Disease.

| Characteristic | Children | Adults | *P* Value |
|---|---|---|---|
| Sex, n (%) | | | .80 |
| Male | 8 (80) | 6 (75) | |
| Female | 2 (20) | 2 (25) | |
| Race, n (%) | | | .37 |
| White | 5 (50) | 6 (75) | |
| Black | 1 (10) | 1 (12.5) | |
| Asian | 2 (20) | 0 | |
| American Indian | 0 | 0 | |
| Other | 2 (20) | 1 (12.5) | |
| Ethnicity, n (%) | | | .80 |
| Not Hispanic | 8 (80) | 6 (75) | |
| Hispanic | 2 (20) | 2 (25) | |
| Laterality, n (%) | | | .25 |
| Unilateral | 10 (100) | 7 (87.5) | |
| Right eye | 3 (30) | 5 (56) | |
| Left eye | 7 (70) | 4 (44) | |
| Bilateral | 0 | 1 (12.5) | |
| Symptom at presentation, n (%) | | | .28 |
| Decreased vision | 5 (50) | 4 (44) | |
| Strabismus | 2 (20) | 0 | |
| No reported symptoms | 3 (30) | 5 (56) | |
| Median (IQR) initial BCVA | 1 (0.54-1.40) | 0 (0-0.32) | <.01[b] |
| Stage[a], n (%) | | | |
| 1 | 0 | 3 (33.3) | .24 |
| 2a | 3 (30) | 3 (33.3) | |
| 2b | 5 (50) | 3 (33.3) | |
| 3a | 1 (10) | 0 | |
| 3b | 1 (10) | 0 | |
| Median (IQR) total clock hours telangiectasia | 2 (1-3) | 1 (1-2) | .10 |
| Median (IQR) total clock hours exudation | 2.5 (2-12) | 1 (0-2) | .01[b] |
| Subretinal fibrosis, n (%) | 2 (20) | 0 | .16 |
| Vitreous hemorrhage, n (%) | 1 (10) | 1 (11) | .94 |
| Vitreoretinal fibrosis, n (%) | 0 | 1 (11) | .28 |

Abbreviation: BCVA, best-corrected visual acuity.
[a]No cases with stage 4 or 5 disease were reported in either group.
[b]Statistically significant.

and fluorescein angiography images from a child and an adult diagnosed with Coats disease.

## *OCTA Findings*

Of the 10 pediatric patients, only 4 patients (40%) had an acceptable quality OCTA image of the affected eye and 2 patients (20%) of the fellow eye, whereas 7 of 8 adult patients (87.5%) had available OCTA of the affected eye and 5 (62.5%) of the fellow eye, all with acceptable quality. Five of the 6 children (83.3%) with poor-quality OCTA images had abnormal OCT findings, including SRF or IRF which, combined with decreased VA, may have contributed to reduced cooperation during imaging, ultimately hindering the acquisition of high-quality images. One adult patient had bilateral disease and was excluded from the comparison with the normal fellow eye. The FAZ area and the vascular density in both the superficial and deep capillary plexi of affected and fellow eyes were analyzed and are described in Supplemental Tables 1 and 2. There was no statistically significant difference in quantitative OCTA metrics between adult and pediatric patients from either the affected or fellow eyes. However, the affected eyes of adult patients with Coats disease had a lower vessel density in most of the quadrants of the superficial and deep capillary plexi compared with their fellow eyes (Supplemental Table 2). This comparison was not made in the pediatric group due to the limited sample size. Qualitative analysis of the 3 × 3 mm OCTA images of the superficial capillary plexus of affected eyes revealed vessels traversing the FAZ in 50% of pediatric patients (Figure 2A), a finding that has previously been reported by Muakkassa et al.[11] Tufts of abnormal neovessels were also seen in 1 child (Figure 2B), and an indistinct FAZ was seen in 1 adult (Figure 2B).

**Table 2.** Comparison of Imaging Findings, Treatment, and Outcomes of Children and Adults With Coats Disease.

| Parameter | Children | Adults | *P* Value |
|---|---|---|---|
| Optical coherence tomography, n (%) | | | |
| Normal | 4 (40) | 8 (89) | .03[b] |
| CME | 0 | 1 (11) | .28 |
| SRF | 4 (40) | 1 (11) | .15 |
| IRF | 2 (20) | 1 (11) | .6 |
| Fluorescein angiography, n (%) | | | |
| Telangiectasias | 9 (90) | 8 (89) | .94 |
| Macular edema | 0 | 1 (11) | .28 |
| Retinal aneurysmal vessels (lightbulb) | 9 (90) | 7 (78) | .47 |
| Perivascular leakage | 8 (80) | 9 (100) | .16 |
| Peripheral capillary nonperfusion | 8 (80) | 8 (89) | .6 |
| Capillary dropout | 4 (40) | 4 (44) | .84 |
| First-line management, n (%) | | | |
| Observation | 0 | 2 (22) | .12 |
| Laser | 4 (40) | 5 (56) | .5 |
| Anti-VEGF | 0 | 1 (11) | .28 |
| Laser and anti-VEGF | 5 (50) | 1 (11) | .07 |
| SRF drainage | 2 (20) | 0 | .16 |
| PPV | 1 (10) | 0 | .16 |
| Mean time to treatment (d) | 13.1 | 30.5 | .49 |
| Retreatment[a] | 6 (60) | 6 (66) | .76 |
| Type of retreatment, n (%) | | | |
| Laser | 5 (83) | 4 (57) | .31 |
| Anti-VEGF | 0 | 2 (29) | .15 |
| Laser and anti-VEGF | 1 (17) | 1 (14) | .91 |
| Mean time to retreatment (d) | 373.00 ± 338.35 | 92.83 ± 123.66 | .04[b] |
| Median (IQR) total number of treatments | 2 (2-4) | 2.5 (1.5-7) | .75 |
| Median (IQR) final BCVA | 0 (0-0.35) | 0.60 (0.54-1.30) | .01 |
| Difference final BCVA between and initial BCVA | −0.07 ± 0.45 | 0.05 ± 0.18 | .59 |
| Median (IQR) length of follow-up (mo) | 37.80 ± 16.19 | 32.94 ± 35.00 | .51 |
| Complications, n (%) | | | |
| None | 4 (40) | 7 (78) | .1 |
| Vitreoretinal fibrosis | 2 (20) | 0 | .16 |
| CME | 1 (10) | 1 (12.5) | .94 |
| Macular scar or atrophy | 4 (40) | 1 (12.5) | .15 |
| ERM | 1 (10) | 1 (12.5) | .94 |
| Cataract | 1 (10) | 1 (12.5) | .94 |
| High IOP | 0 | 1 (12.5) | .28 |
| Corneal opacity | 0 | 0 | — |
| NVG | 0 | 0 | — |
| NVI | 0 | 0 | — |
| Phthisis bulbi | 0 | 0 | — |

Abbreviations: Anti-VEGF, antivascular endothelial growth factor; BCVA, best-corrected visual acuity; CME, cystoid macular edema; ERM, epiretinal membrane; IOP, intraocular pressure; IRF, intraretinal fluid; NVI, neovascularization of the iris; NVG, neovascular glaucoma; PPV, pars plana vitrectomy; SRF, subretinal fluid.

[a]Indications for retreatment included lack of improvement/persistence, progression, and/or recurrence of the disease's retinal features.

[b]Statistically significant.

## *Treatment and Outcomes*

Treatment and outcomes are summarized in Table 2. The most common first-line treatment in children was combined laser photocoagulation and intravitreal (IVT) antivascular endothelial growth factor (anti-VEGF) injection (50%), while in adults, the most common first-line treatment was laser only (56%). Vitrectomy (20%) and SRF drainage (20%) were performed only in children. Two adults (22%) were observed without receiving any treatment. The median total number of treatments received in children was 2.5 and was 2 in adults. Although the change in VA during follow-up visits did not differ between either group, the VA at final follow-up was significantly lower in children (median, 0.60; IQR, 0.54-1.30, or Snellen 20/80) than adults (median logMAR, 0; IQR, 0-0.35, or Snellen 20/20) ($P$ = .01). Although the rates of some sequelae were similar between

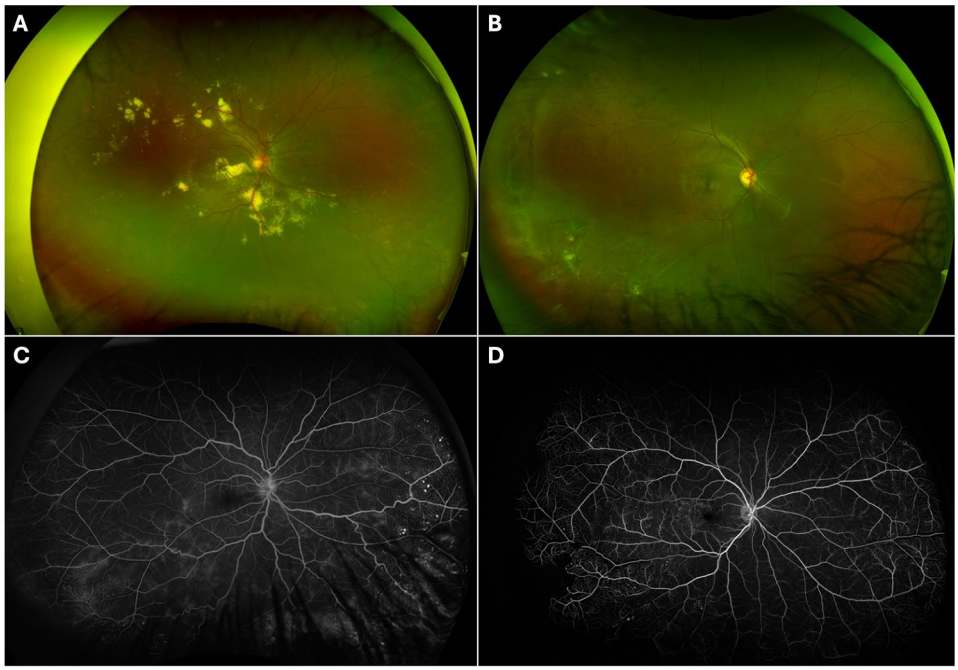


**Figure 1.** (A) Fundus photograph of the right eye of a child with Coats disease shows diffuse yellow exudates limited to an area within the equator, with a radial spoke surrounding the macula. (C) Corresponding fluorescein angiography shows telangiectatic vessels and aneurysms in the retinal periphery, mostly nasally and inferiorly. (B) Fundus photograph of the right eye of an adult with Coats disease shows peripheral sclerotic vessels temporally with capillary dropout in the far temporal periphery and chorioretinal atrophy temporally and inferotemporally. (D) Corresponding fluorescein angiography shows peripheral telangiectasia and nonperfusion, with several pinpoint spots of hyperfluorescence in the periphery.

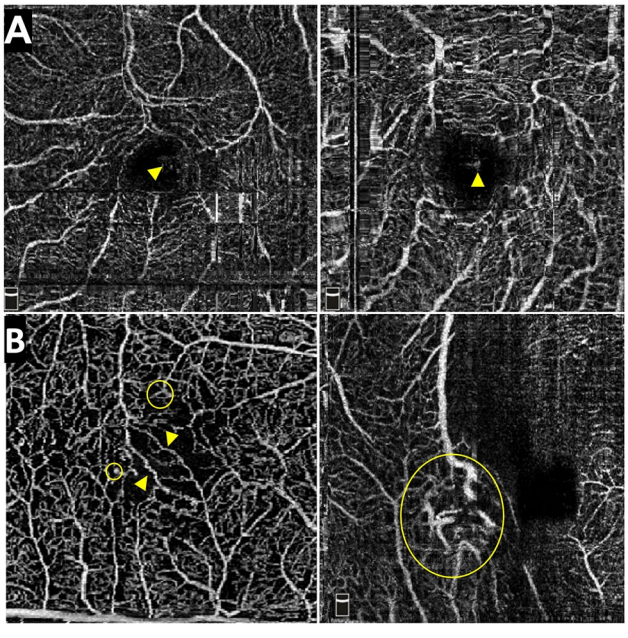


**Figure 2.** Qualitative analysis of 3 × 3-mm optical coherence tomography angiography images of the superficial plexus in (A) children and (B) adults shows vessels traversing the foveal avascular zone (arrowheads), indistinct foveal avascular zone (B), and tufts of abnormal neovessels (circle in B).

each group, a higher proportion of pediatric vs adult patients presented with macula fibrosis (40% vs 12.5%), of whom 75% vs 0% had macular involvement.

## Conclusions

To our knowledge, this is the first study to compare quantitative and qualitative OCTA findings, in addition to clinical features, between adult and pediatric populations with Coats disease. OCTA images show flow in the retinal and choroidal vasculature, and it has been valuable to visualize anomalous vasculature in other disorders of the posterior pole. Our findings demonstrated that there was a greater number of qualitative OCTA findings in children with Coats disease, including vessels traversing the FAZ, tufts of abnormal neovessels, and indistinct FAZ than adults. OCTA is very sensitive to loss of fixation and motion artifact, making it more challenging for images to be taken in the pediatric age group, especially if poor vision limits fixation. It was also noted that adult patients had a decrease in vessel density in the affected eye compared with the fellow eye.

### *Role of OCTA in the Diagnosis of Adult Versus Pediatric Coats Disease*

In recent years, OCTA has been used noninvasively to investigate pediatric retinal diseases, providing high-resolution and

depth-resolution images. Previous research in pediatric patients with Coats disease suggested a decrease in vessel density in both the superficial and deep capillary plexi as well as an enlargement of the FAZ.[14] Ours is the first study to show a lower vessel density of the superficial capillary plexus and deep capillary plexus in the affected eyes of adult patients compared with their fellow eyes. These microvascular alterations within the macula may be a secondary result of the peripheral pathology. There were no differences found between adult and pediatric quantitative metrics on OCTA, but this may be due to sample size. It is also noteworthy that the literature lacks a normative database of OCTA metrics in the pediatric population that would allow baseline characteristics to be compared between adults and children.

The qualitative OCTA findings, including an indistinct FAZ, vessels traversing the FAZ, and tufts of abnormal neovessels, were more common in pediatric patients. They were more commonly found in subjects with a stage 2b or higher disease. Of note, these vascular abnormalities of the macula were not seen on FA, likely due to its limited resolution in detecting subtle microvascular changes within the FAZ. Unlike OCTA, FA highlights perfused vessels but may miss nonleaking structural anomalies such as capillary remodeling. Our findings indicate that OCTA may provide better detection of macular vascular abnormalities in patients with Coats disease,[11,19] possibly because of the unique ability of OCTA to segment 3 distinct retinal capillary plexuses,[20] making it a sensitive device for the detection of early subclinical central vascular changes.[14]

### *OCT and FA in Adult Versus Pediatric Coats Disease*

Multimodal imaging, including fundus imaging, FA, and OCT, are used to characterize the clinical features associated with Coats disease.[12] FA is considered the standard for revealing hyperfluorescence, highlighting the telangiectatic vessels, perivascular leakage, aneurysms, capillary dropout, and peripheral capillary nonperfusion. These angiographic parameters were similarly represented between adults and children as common diagnostic features for both age groups. However, more abnormalities were seen in children on macular OCT B-scan, including SRF and IRF, compared with adults ($P = .03$). This finding was expected because pediatric patients presented with more advanced disease involving the macula (70%) than adults, whose disease spared the fovea in two-thirds of the cases. A more frequent foveal involvement in younger vs older patients was also reported by Dalvin et al[8] (98% vs 77%, $P = .002$).

### *Characteristics and Severity of Coats Disease in Adult Versus Children*

Although Coats disease is unilateral in more than 95% of cases,[21] previous studies have shown that bilateral involvement can occur, even in the absence of visible clinical abnormalities, which prompts a complete evaluation of the fellow eye.[10,12,22,23] Most patients in the current study had unilateral disease (94.4%) with normal OCT and FA images of the fellow eyes.

Daruich et al[7] showed that earlier onset of Coats disease in the pediatric group is associated with more severe manifestations, advanced stages, and worse visual outcome, confirming findings from previous smaller reports.[5,6] Similarly, Dalvin et al[8] concluded that younger patients with Coats disease present with worse VA, more advanced stages of disease, and are more likely to require enucleation as primary treatment because of disease severity at presentation, rather than disease progression over time. Our study results confirm these findings, with children presenting with a significantly lower initial VA ($P < .01$), greater total clock hours of exudation ($P = .01$), advanced stage, and more macula OCT findings ($P = .03$). Feng et al[24] found that a higher concentration of inflammatory markers in children, such as VEGF, interleukin-6, and interleukin-1β, as compared with adults with Coats disease may play a role in differences in disease severity. In addition, pediatric patients may be less likely to report reduced VA, which may lead to a delayed presentation compared with adults.

### *Treatment of Coats Disease in Adults Versus Children*

Although combined laser photocoagulation and IVT anti-VEGF injection was the most common first-line treatment in children, laser only was employed in most of the adult cases, with 22% managed by observation only. These numbers reflect the tendency toward a more aggressive approach in managing younger patients with more severe disease. Finally, the VA at final follow-up was significantly lower in children compared with adults, as seen in previous reports.[8,25] This result was expected because children presented with worse vision and more severe disease with more macular involvement. Of note, amblyopia might have contributed to the decrease in the VA in these pediatric patients.

Overall, our study confirmed that children present with more advanced disease, with OCTA showing differences between adults and pediatric patients. There was a greater number of qualitative OCTA findings in children, including vessels traversing the FAZ, tufts of abnormal neovessels, and an indistinct FAZ. In adults, there was a decrease in vessel density in the affected eye compared with the fellow eye.

Limitations of the current study include its retrospective nature and a small sample size, which may reduce the generalizability of the findings. The number of pediatric OCTA images for quantitative analysis of vessel density and the FAZ was limited due to motion artifact, poor fixation, and cooperation issues, which underscores the challenges of performing OCTA in younger children who may not cooperate well during testing.

In conclusion, younger age is associated with more advanced Coats disease at presentation, leading to worse prognosis. Similar quantitative metrics between children and adult patients

were seen with OCTA, but there were more qualitative abnormalities in the pediatric group, specifically in the superficial vascular plexus. Future studies with larger samples will be crucial to further characterize the use of multimodal imaging in both pediatric and adult populations.

## Ethical Approval

This study was conducted in accordance with the Declaration of Helsinki. The collection and evaluation of all protected patient health information were performed in a US Health Insurance Portability and Accountability Act–compliant manner. This study was approved by the Institutional Review Board of Massachusetts General Brigham (protocol ID# 2021P002833).

## Statement of Informed Consent

The requirement for informed consent was waived given the retrospective nature of the study.

## Declaration of Conflicting Interests

The author(s) declared the following potential conflicts of interest with respect to the research, authorship, and/or publication of the article: Dr. Patel is a consultant to Alcon, Alimera, Allergan, Apellis, Atheneum, Biogen, Dorc, EyePoint, Genentech, Gerson Lehrman Group, Inc, Guidepoint, Kyoto Drug Company, Lifesciences, RegenxBio, and Regeneron. Dr. Baumal is the chief medical officer for Apellis. None of the other authors declared potential conflicts of interest with respect to the research, authorship, and/or publication of the article.

## Funding

Dr. Hoyek is supported by the VitreoRetinal Surgery Foundation. Dr. Patel is supported by the Retina Innovation Fund, Massachusetts Eye and Ear, Boston, MA, USA. Dr. Mukai is supported in part by gifts to the Mukai Fund, Massachusetts Eye and Ear, Boston, MA, USA. The funding organizations had no role in design or conduct of this research.

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## Supplementary Material

Supplementary material is available online with this article.

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