American Journal of Ophthalmology Case Reports 37 (2025) 102232

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

Coats disease presenting with vitreous hemorrhage and neovascular glaucoma

Amrish Selvam, Hesham Gabr, R.V. Paul Chan, Michael J. Heiferman *

Department of Ophthalmology and Visual Sciences, University of Illinois at Chicago, Chicago, 60612, USA

ARTICLE INFO

Keywords:
Coats disease
Neovascular glaucoma
Vitreous hemorrhage
Vitrectomy

ABSTRACT

Purpose: To describe a patient with Coats disease with an atypical presentation of neovascular glaucoma and vitreous hemorrhage.

Observations: A 15-year-old male presented with five days of pain, redness, and swelling and was found to have neovascular glaucoma in his right eye. Further evaluation revealed Coats disease stage 3AI with a subtotal exudative retinal detachment inferiorly, telangiectatic vessels, and vitreous hemorrhage. Intravitreal anti-vascular endothelial growth factor, laser photocoagulation, pars plana vitrectomy, and valved tube-shunt placement was performed, eventually resulting in a favorable visual outcome.

Conclusions and importance: We report a patient with a unique presentation of advanced Coats disease. By highlighting the treatment approach and potential factors that led to the preservation of visual function, we hope to expand the literature on the diagnosis and management of Coats disease.

1. Introduction

Coats disease is a vascular disorder characterized by retinal telangiectasia and aneurysms with exudation.[1–3] The leaky telangiectatic vessels lead to intraretinal and subretinal exudation with outer retinal thickening. Partial or total retinal detachment without retinal traction may subsequently follow. If left untreated, Coats disease can cause blindness and lead to enucleation.[4,5] Therefore, urgent diagnosis and management of Coats disease is critical in preserving visual function.

Coats disease usually affects unilateral eyes of young males in their first two decades of life with an incidence of 0.09 per 100,000.[6] In a multi-center study of Coats disease, angiographic changes affecting the fellow eye were found in 18.8% of cases, although these lesions were not clinically significant and did not progress.[7] Adult onset of the disease has also been reported although with a slower rate of progression and localized lipid deposition.[8]

Common clinical signs or symptoms include decreased visual acuity, strabismus, leukocoria, and pain 2. Most patients present with a normal anterior segment although a minority present with neovascular glaucoma (NVG), corneal edema, neovascularization of the iris, and megalocornea.[2,9]

The diagnosis of Coats Disease is made clinically using indirect ophthalmoscopy, but fluorescein angiography (FA), optical coherence tomography (OCT), and ultrasound provide good utility.[2,10,11] On FA, areas of non-perfusion, early hyperfluorescence of telangiectasia that persists to the late stage, and exudation hyperfluorescence can indicate the presence of Coats disease.[3,12] Telangiectasias and aneurysms are seen as “light bulb” dilations due to the bulbous configuration of the vessels and surrounding yellow exudation.[3,9,13]

In this report, we discuss a unique presentation of Coats disease with NVG and vitreous hemorrhage.

2. Case presentation

A 15-year-old Hispanic male presented with five days of progressively worsening sudden-onset pain in his right eye upon awakening associated with eye redness and blurry vision. He had no significant prior ocular history, pertinent medical history, or recent trauma. This was his first experience with such an episode.

On examination of the right eye, he was found to have an intraocular pressure (IOP) of 53 mmHg, visual acuity of count fingers at three inches, a mid-dilated but reactive pupil, conjunctival injection, diffuse microcystic epithelial corneal edema, layered hyphema, and iris neovascularization at the pupillary margin in the right eye. Gonioscopy revealed peripheral anterior synechia in all quadrants of the right eye. Posterior segment exam was limited by the anterior findings. Ultrasound

* Corresponding author.1009 S Wood St, Chicago, IL, 60612, USA.
E-mail address: mheif@uic.edu (M.J. Heiferman).

https://doi.org/10.1016/j.ajoc.2024.102232
Received 6 September 2024; Received in revised form 18 November 2024; Accepted 7 December 2024
Available online 13 December 2024

revealed vitreous opacities and an inferior retinal detachment without any obvious retinal tears, posterior masses, or calcifications. Exam findings in the left eye were within normal limits.

Given these observations, the patient was diagnosed with NVG and started on medical management with the goal of controlling the elevated IOP and improving the corneal edema to achieve a better view of the posterior segment. The patient was started on topical carbonic anhydrase inhibitor and beta blocker (dorzolamide/timolol) twice a day in the right eye, systemic carbonic anhydrase inhibitor (acetazolamide 500 mg) twice a day in the right eye, topical steroid (prednisolone acetate 1%) six times a day in the right eye, and cyclopentolate 1% twice a day in the right eye with close follow up.

Over the following week, the patient observed improvement in visual acuity (20/100), IOP (30 mmHg), and symptoms. Resolution of hyphema and reduction in corneal edema allowed visualization of the posterior segment which showed vitreous hemorrhage and exudative retinal detachment inferiorly extending inferotemporally and inferonasally sparing the macula. FA showed diffuse vascular leakage inferiorly along telangiectatic vessels with "light bulb" dilations as seen in Fig. 1, suggestive of Coats Disease.

Inflammatory and infectious workup was obtained to evaluate for the underlying etiology including negative Quantiferon, syphilis, lysozyme, angiotensin converting enzyme, and chest x-ray. Given the negative workup, the diagnosis of Coats Disease was made. With an IOP ranging from 30 to 45 mmHg, netarsudil 0.02% twice a day in the right eye was added.

Two weeks after the initial presentation, the patient reported an improvement in symptoms with a visual acuity of 20/60. IOP improved but remained elevated >25 mmHg likely due to peripheral anterior synechia. The patient underwent an examination under anesthesia with laser photocoagulation to the observed areas of exudation and telangiectatic vessels inferiorly using a laser indirect ophthalmoscope (LIO) which was limited by the vitreous hemorrhage. An intravitreal injection of anti-vascular endothelial growth factor (anti-VEGF), bevacizumab 1.25mg/0.05 mL, was performed in the right eye given the NVG.

One week following laser and anti-VEGF therapy, elevated IOP >30 mmHg and vitreous hemorrhage were still present. Given the persistent elevation of the IOP and vitreous hemorrhage the decision was made to proceed with a tube-shunt insertion in the anterior chamber and pars plana vitrectomy with endolaser in the right eye. Adequate endolaser uptake was observed at the lightbulb aneurysms and telangiectatic vessels.

Post-operative visits showed improvement in IOP (10 mmHg), improved subretinal fluid, and clumping of vitreous heme inferiorly by post-operative day 4 in the right eye. The patient was tapered off prednisolone and medications were reduced to only topical dorzolamide/timolol twice a day in the right eye. Imaging performed during this post-operative period are shown in Fig. 2.

Three months following surgery, FA showed persistent vascular leakage despite clinical improvement prompting additional laser photocoagulation targeting abnormal blood vessels in the affected area in the right eye using an LIO under general anesthesia. The exudative retinal detachment resolved and the patient was monitored closely for the following 18 months. Best corrected visual acuity improved to 20/40 and IOP remained <12 for subsequent visits. Follow up FAs showed no vascular leakage. Management was continued with topical dorzolamide/timolol twice a day in the right eye. Imaging taken after the final round of laser but prior to the resolution of the exudative retinal detachment is shown in Fig. 3.

3. Discussion

Coats disease is a telangiectatic retinal disease that carries a poor prognosis, particularly when presenting with NVG and vitreous hemorrhage, as seen in this patient. According to the Shield et al. classification, this patient had stage 3AI Coats disease with an extrafoveal subtotal detachment.[14] In brief, stage 1 of the Shield et al. classification of Coats disease is characterized by telangiectasia only, stage 2 by telangiectasia with (A) extrafoveal exudation or (B) foveal exudation, and stage 3 by exudative retinal detachment. Stage 3 can be further classified into (A) subtotal retinal detachment that is (I) extrafoveal or (II) foveal or (B) total retinal detachment. Stage 4 is characterized by total retinal detachment with glaucoma, and stage 5 by advanced end-stage disease.[14] Visual acuity after treatment in this stage is 20/40 or better in 42% of patients and worse than 20/200 in 31% of patients, with the primary causes of vision loss being retinal detachment and macular scarring.[15] In the presented case, laser photocoagulation and anti-VEGF therapy alone did not successfully control the IOP due to the presence of NVG with peripheral anterior synechia and the incomplete initial laser treatment given the vitreous hemorrhage. However, after tube-shunt insertion and pars plana vitrectomy with endolaser, the patient experienced significant improvement in symptoms with objective improvement in his vision and IOP. A final round of laser photocoagulation led to better control of the disease, stable vision and IOP, and resolution of leakage on FA. This case highlights the management of complications associated with Coats disease and the utility of serial FAs in the diagnosis and monitoring of the disease.

Neovascularization of the iris and NVG are poor prognostic factors more commonly associated with stage 3B, 4, and 5 Coats disease.[15] In the past, NVG in Coats disease was treated with primary enucleation to

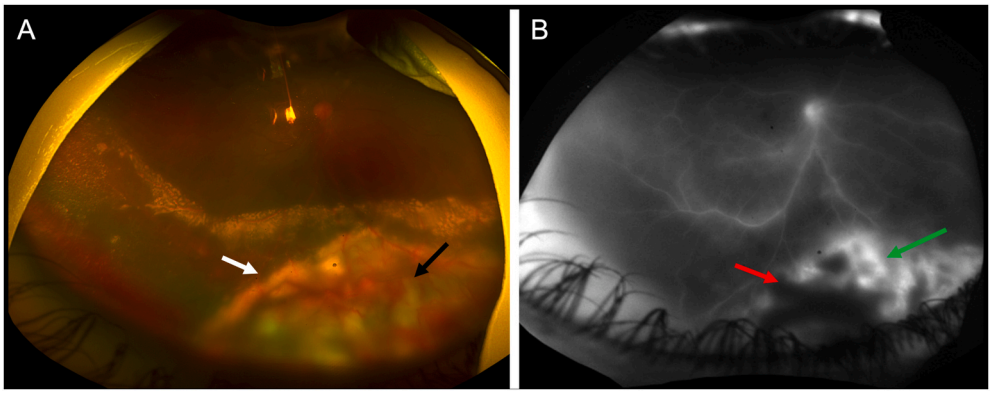

Fig. 1. Ophthalmic retinal imaging of a 15-year-old male newly diagnosed with Coats disease in the right eye presenting with five days of ocular pain. Images were acquired after the resolution of the corneal edema but were limited by the presence of vitreous hemorrhage. (A) Ultra-widefield fundus image with telangiectatic vessels (black arrow) and exudative retinal detachment inferiorly (white arrow). (B) Fluorescein angiography in the late-phase showing vascular leakage (green arrow), "light bulb" dilations, and areas of nonperfusion (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

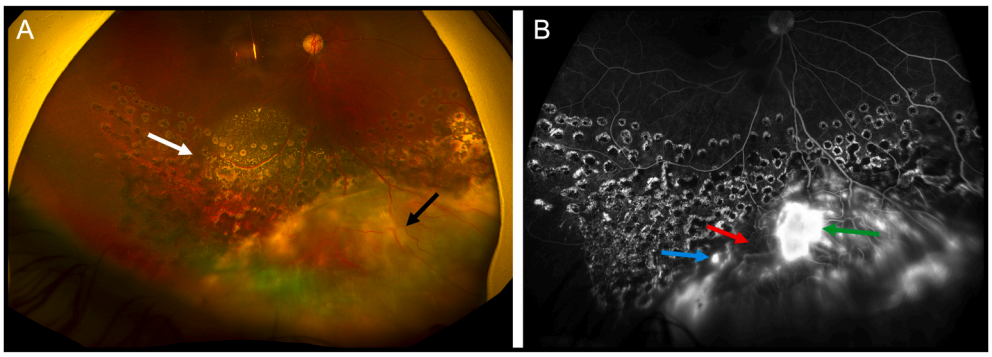

Fig. 2. Ophthalmic retinal imaging of the right eye in a 15-year-old male with Coats disease taken during the post-operative period following tube-shunt insertion and pars plana vitrectomy. (A) Fundus imaging centered inferiorly shows diffuse photocoagulation scarring from retinal laser (white arrow), improvement in the subretinal fluid, and telangiectatic vessels (black arrow). (B) Fluorescein angiography in the arteriovenous phase shows persistent vascular leakage (green arrow), capillary nonperfusion (red arrow), and "light bulb" dilations (blue arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

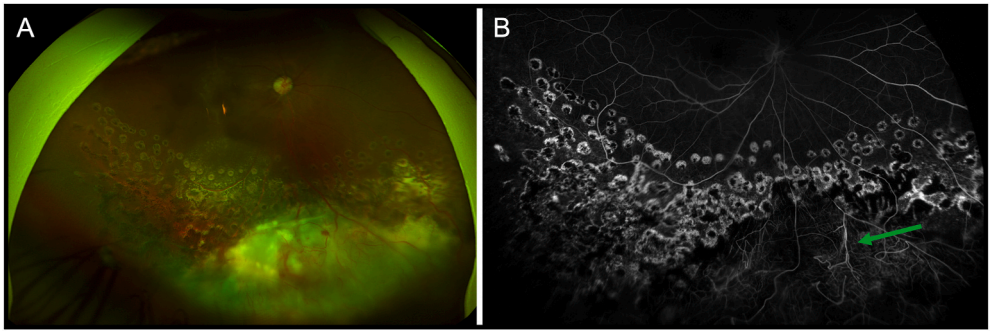

Fig. 3. Ophthalmic retinal imaging of the right eye in a 15-year-old male with Coats disease taken 1 month after additional laser photocoagulation and 4 months after pars plana vitrectomy. (A) Fundus imaging shows diffuse scarring from photocoagulation with chronic exudative retinal detachment inferiorly unchanged from prior imaging. (B) Venous stage fluorescein angiography showing reduced vascular leakage in areas affected by Coats disease (green arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

relieve severe ocular pain. In a study by Shields et al., in 2001, all patients with corneal edema, and the majority of patients with neovascularization of the iris or ocular hypertension received enucleation.[2] The presence of NVG alone is a primary predictor of enucleation in patients with Coats disease.[2,9] For instance, a case of a 2-year-old child with facioscapulohumeral dystrophy was found to have Coats disease bilaterally, but only required enucleation of the right eye where NVG was present.[16] However, earlier diagnosis and improvements in NVG treatment have led to the preservation of vision using other treatment modalities such as intravitreal anti-VEGF therapy.[2,17] Since NVG alters the anatomy of the iridocorneal angle by proliferated tissues, surgical intervention may be necessary to achieve target IOP. In this case, the patient saw improvement with the placement of a valved tube-shunt drainage device. These devices have shown great efficacy in refractory glaucoma, and as this case demonstrates, may be an effective means of preserving visual function in patients with NVG secondary to Coats disease.[18,19]

Vitreous hemorrhage is a rare finding of Coats disease. Typically, aneurysms in Coats disease do not bleed and are more commonly associated with exudative retinal detachment.[20] Despite FA showing capillary nonperfusion in areas surrounding the telangiectatic vessels, neovascularization is rarely observed, which may also explain the rarity of vitreous hemorrhage in these eyes. Instead, vitreous hemorrhage may result from neovascularization secondary to chronic retinal detachments.[9] Of the 158 eyes with Coats disease reported by Shields et al., in 2001, none reported vitreous hemorrhage.[2] In an Indian cohort, Rishi et al. found that 4.3% of eyes presented with vitreous hemorrhage that precluded a complete posterior segment exam.[21] Shanmugan et al. evaluated patients with advanced Coats disease and found a higher prevalence of vitreous hemorrhage, 31.3% in advanced and 7.7% in stage 3A, which was associated with poor functional and anatomic outcomes.[22] While pars plana vitrectomy is not often indicated in patients with stage 3AI, it was used for our patient to remove the non-clearing vitreous hemorrhage and ensure a timely and complete laser photocoagulation to the affected areas. Since the patient's macula and central vision were largely unaffected by pathology, he had good visual potential. Therefore, pars plana vitrectomy should be considered in patients with Coats disease who have a limited view for laser photocoagulation but good visual potential.

Due to NVG, this patient's initial diagnosis of Coats disease was challenging as examination of the posterior segment was limited by corneal edema, hyphema, and vitreous hemorrhage. Multimodal imaging was used throughout the management of this patient. Ultrasound played an important role in filling these gaps and revealing the vitreous hemorrhage and exudative retinal detachment without any masses. FA showed telangiectatic vessels and light bulb aneurysms to confirm the diagnosis, and repeat FAs were used to monitor for resolution of the vascular leakage to guide post-operative management. Recent literature supports the routine use of wide-angle FA and FA-guided treatment in the management of Coats disease, which can result in outstanding visual and anatomic outcomes.[13,23] In addition, the use of FA imaging may allow clinicians to catch early-stage Coats disease in asymptomatic patients and fellow eyes.[24]

Demographics are also a factor when considering the prognosis of Coats disease. Older age is associated with a more favorable prognosis.[25] The adult variety often presents asymptomatically and may be picked up

by routine retinal findings. While our patient is not an adult, his older age relative to the median for Coats disease is consistent with better visual outcomes as observed in other studies.[4,25] One study evaluated the visual outcomes in a small cohort of Hispanic patients and found that Coats disease presented at an earlier age with macular symptoms and poor visual outcomes.[26] However, this was not the case for our patient. Given this atypical presentation of Coats disease, a secondary Coats-like reaction was also considered. Our patient had no evidence of inherited dystrophies or retinal vasculitis and no medical problems to suggest a systemic association or genetic condition. Secondary Coats-like reactions can have a worse prognosis compared to pediatric Coats disease making early identification of the primary etiology important for patient counseling and management decisions.[27]

4. Conclusion

NVG is associated with a poor prognosis and vitreous hemorrhage is an atypical finding in Coats disease. Despite these complications, our patient achieved a good visual outcome after vitrectomy, tube-shunt placement, and multiple sessions of laser photocoagulation guided by FA. Despite a favorable visual outcome in this case, much is still unknown regarding the etiology of Coats disease and various presenting features. Further research is necessary to better understand the disease course and provide personalized treatment options.

CRediT authorship contribution statement

Amrish Selvam: Writing – original draft, Visualization, Investigation, Formal analysis, Data curation. **Hesham Gabr:** Writing – review & editing, Supervision, Conceptualization. **R.V. Paul Chan:** Writing – review & editing, Conceptualization. **Michael J. Heiferman:** Writing – review & editing, Supervision, Resources, Conceptualization.

Disclosures

None.

Funding

This research was supported by an unrestricted departmental grant from Research to Prevent Blindness, New York, NY, National Eye Institute P30 EY001792, and National Eye Institute K12 EY021475.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Acknowledgments

None.

References

1. Coats G. Forms of retinal disease with massive exudation. *Roy London Ophthalmol Hosp Rep*. 1908;17:440–525.
2. Shields JA, Shields CL, Honavar SG, Demirci H. Clinical variations and complications of Coats disease in 150 cases: the 2000 sanford gifford memorial lecture. *Am J Ophthalmol*. 2001;131(5):561–571. https://doi.org/10.1016/S0002-9394(00)00883-7, 2001/05/01.
3. Gupta A, Paulbuddhe VS, Shukla UV, Tripathy K. Exudative retinitis (Coats disease). *StatPearls*. 2024. Treasure Island (FL).
4. Daruich A, Matet A, Munier FL. Younger age at presentation in children with coats DISease IS associated with more advanced stage and worse VISual prognosIS: a retrospective study. *Retina*. 2018;38(11):2239–2246. https://doi.org/10.1097/iae.0000000000001866.
5. Ruben M, Yaghy A, Park CH, Shields CL. Neglected Coats disease. *J Pediatr Ophthalmol Strabismus*. Oct 20 2020;57:e82–e85. https://doi.org/10.3928/01913913-20200818-01 (in eng).
6. Morris B, Foot B, Mulvihill A. A population-based study of Coats disease in the United Kingdom I: epidemiology and clinical features at diagnosis. *Eye*. Dec 2010;24(12):1797–1801. https://doi.org/10.1038/eye.2010.1801.
7. Jeng-Miller KW, Soomro T, Scott NL, et al. Longitudinal examination of fellow-eye vascular anomalies in Coats' disease with widefield fluorescein angiography: a multicenter study. *Ophthalmic Surg Lasers Imaging Retina*. Apr 1 2019;50(4):221–227. https://doi.org/10.3928/23258160-20190401-04 (in eng).
8. Banerjee M, Nayak S, Kumar S, Bhayana AA, Kumar V. Adult-onset Coats disease. *Surv Ophthalmol*. 2023/07/01/2023;68(4):591–600. https://doi.org/10.1016/j.survophthal.2023.03.002.
9. Sen M, Shields CL, Honavar SG, Shields JA. Coats disease: an overview of classification, management and outcomes. *Indian J Ophthalmol*. Jun 2019;67(6):763–771. https://doi.org/10.4103/ijo.IJO_841_19 (in eng).
10. Spitznas M, Joussen F, Wessing A, Meyer-Schwickerath G. Coat's disease. An epidemiologic and Fluorescein angiographic study. *Albrecht Von Graefes Arch Klin Exp Ophthalmol*. Jul 4 1975;195(4):241–250. https://doi.org/10.1007/bf00414937 (in eng).
11. Adam RS, Kertes PJ, Lam WC. Observations on the management of Coats' disease: less is more. *Br J Ophthalmol*. Mar 2007;91(3):303–306. https://doi.org/10.1136/bjo.2006.103382 (in eng).
12. Ruia S, Tripathy K. Fluorescein angiography. In: *StatPearls. Treasure Island (FL)*. 2024.
13. Yang X, Wang C, Su G. Recent advances in the diagnosis and treatment of Coats' disease. *Int Ophthalmol*. 2019/04/01 2019;39(4):957–970. https://doi.org/10.1007/s10792-019-01095-8.
14. Shields JA, Shields CL, Honavar SG, Demirci H, Cater J. Classification and management of Coats disease: the 2000 proctor lecture. *Am J Ophthalmol*. May 2001;131(5):572–583. https://doi.org/10.1016/s0002-9394(01)00896-0 (in eng).
15. Shields CL, Udyaver S, Dalvin LA, et al. Visual acuity outcomes in Coats disease by classification stage in 160 patients. *Br J Ophthalmol*. 2020;104(3):422. https://doi.org/10.1136/bjophthalmol-2019-314363.
16. Shields CL, Zahler J, Falk N, et al. Neovascular glaucoma from advanced Coats disease as the initial manifestation of facioscapulohumeral dystrophy in a 2-year-old child. *Arch Ophthalmol*. 2007;125(6):840–842. https://doi.org/10.1001/archopht.125.6.840.
17. de Silva DJ, Brookes JL. Cyclodiode treatment of neovascular glaucoma secondary to Coats' disease. *Br J Ophthalmol*. May 2007;91(5):690–691. https://doi.org/10.1136/bjo.2006.097121 (in eng).
18. Dumbrăveanu L, Cușnir V, Bobescu D. A review of neovascular glaucoma. Etiopathogenesis and treatment. *Rom J Ophthalmol*. Oct-Dec 2021;65(4):315–329. https://doi.org/10.22336/rjo.2021.66 (in eng).
19. Shalaby WS, Ganjei AY, Wogu B, et al. Outcomes of Ahmed glaucoma valve and transscleral cyclophotocoagulation in neovascular glaucoma. *Indian J Ophthalmol*. Apr 2022;70(4):1253–1259. https://doi.org/10.4103/ijo.IJO_2107_21 (in eng).
20. Grosso A, Pellegrini M, Cereda MG, Panico C, Staurenghi G, Sigler EJ. Pearls and pitfalls in diagnosis and management of coats disease. *Retina*. 2015;35(4) [Online]. Available: https://journals.lww.com/retinajournal/fulltext/2015/04000/pearls_and_pitfalls_in_diagnosis_and_management_of.2.aspx.
21. Rishi P, Rishi E, Uparkar M, et al. Coats' disease: an Indian perspective. *Indian J Ophthalmol*. Mar-Apr 2010;58(2):119–124. https://doi.org/10.4103/0301-4738.60081 (in eng).
22. Shanmugam MP, Shah PN, Sagar P, Ramanjulu R, Mishra DK. Coats' disease - prognostic factors for globe and vision salvage in children, a long-term experience. *Indian J Ophthalmol*. Feb 2022;70(2):483–489. https://doi.org/10.4103/ijo.IJO_1822_21 (in eng).
23. Suzani M, Moore AT. Intraoperative fluorescein angiography-guided treatment in children with early Coats' disease. *Ophthalmology*. Jun 2015;122(6):1195–1202. https://doi.org/10.1016/j.ophtha.2015.02.002 (in eng).
24. Jung EH, Kim JH, Kim SJ, Yu YS. Fluorescein angiographic abnormalities in the contralateral eye with normal fundus in children with unilateral Coats' disease. *Kor J Ophthalmol*. Feb 2018;32(1):65–69. https://doi.org/10.3341/kjo.2016.0092 (in eng).
25. Dalvin LA, Udyaver S, Lim LS, et al. Coats disease: clinical features and outcomes by age category in 351 cases. *J Pediatr Ophthalmol Strabismus*. Sep 1 2019;56(5):288–296. https://doi.org/10.3928/01913913-20190716-01 (in eng).
26. Barrera-Sanchez M, Martinez-Munoz RE, Ruiz-Lozano RE, Busch C, Paez-Garza JH, Rodriguez-Valdes PJ. Coats disease in 9 patients: a hispanic case series. *Klin Monbl Augenheilkd*. Jul 2022;239(7):923–928. https://doi.org/10.1055/a-1658-0772. Morbus Coats in 9 Patienten: eine hispanische Fallserie.
27. Nieves FR, Villegas VM, Patel NA, Berrocal AM, Murray TG. Multimodal treatment of Coats-like exudative vitreoretinopathy in Goldmann-Favre syndrome. *Am J Ophthalmol Case Rep*. Mar 2022;25, 101362. https://doi.org/10.1016/j.ajoc.2022.101362 (in eng).