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Comparison of Visual Outcomes in Coats' Disease

A 20-Year Experience

Sally S. Ong, MD,[1] *Edward G. Buckley, MD,*[1] *Brooks W. McCuen II, MD,*[1] *Glenn J. Jaffe, MD,*[1] *Eric A. Postel, MD,*[1] *Tamer H. Mahmoud, MD, PhD,*[1] *Sandra S. Stinnett, DrPH,*[1] *Cynthia A. Toth, MD,*[1] *Lejla Vajzovic, MD,*[1] *Prithvi Mruthyunjaya, MD, MHS*[1,2]

Purpose: To report differences in visual acuities among patients with Coats' disease who sought treatment at a tertiary care university-based practice.

Design: Single-center retrospective cohort study.

Participants: Patients with Coats' disease diagnosed clinically, angiographically, or both from 1995 through 2015.

Methods: Patients were divided into 2 groups based on date of presentation: decade 1 (1995–2005) and decade 2 (2006–2015).

Main Outcome Measures: Visual acuity (VA).

Results: Thirty-nine eyes of 39 patients were included with 19 eyes presenting in decade 1 and 20 eyes presenting in decade 2. Three patients demonstrated bilateral disease, but only the worse eye was included for analysis. Forty-seven percent of eyes in decade 1 demonstrated advanced stages of disease (stage 3B or worse) compared with 20% of eyes in decade 2. There was a trend for the mean initial presenting VA (±standard deviation) for decade 1 eyes to be worse (2.05±1.29 logarithm of the minimum angle of resolution [logMAR]) than for decade 2 eyes (1.45±0.99 logMAR; $P = 0.1$). From initial to final follow-up visit, mean VA also worsened for decade 1 eyes ($P = 0.03$), but remained stable for decade 2 eyes ($P = 1.0$). At the end of follow-up, there was a trend for mean VA for decade 1 eyes (2.28±1.17 logMAR) to be worse than for decade 2 eyes (1.60±1.15 logMAR; $P = 0.07$). Eight eyes were observed initially in decade 1 compared with 1 eye in decade 2, and only 1 of the observed eyes (in decade 2) developed painful glaucoma requiring enucleation. Decade 2 eyes had a higher average number of procedures per eye (6.5±4.9) compared with decade 1 eyes (1.4±1.7; $P < 0.001$).

Conclusions: The earlier presentation of disease in decade 2 suggests improvements in disease detection over time. Furthermore, there was a trend for eyes to have better final VA in this decade. This is due to a combination of factors, including earlier presentation of disease, fewer eyes being observed without treatment, and eyes, when treated, receiving a higher number of procedures. *Ophthalmology 2017;■:1–9 © 2017 by the American Academy of Ophthalmology*

Coats' disease is a sporadic and nonhereditary condition characterized as idiopathic retinal telangiectasia that may progress to retinal exudation and exudative retinal detachment in the absence of retinal or vitreous traction.[1] Severe complications of untreated Coats' disease can be devastating and include neovascular glaucoma and phthisis bulbi.[2]

Traditionally, Coats' disease has been diagnosed clinically with indirect ophthalmoscopy, and in eyes with early stages of disease, has been treated with laser photocoagulation or cryotherapy to the abnormal telangiectasic vessels.[1] In recent years, the advent of wide-field imaging and fluorescein angiography during examination under anesthesia has allowed earlier identification of peripheral vascular anomalies and areas of abnormal retinal perfusion in one or both eyes.[3–5] In cases where subretinal fluid prevents adequate ablative therapy, Coats' disease has been treated with intravitreal triamcinolone (IVT) or anti–vascular endothelial growth factor (VEGF) injections, or surgically with vitrectomy, subretinal fluid drainage, or both, with or without scleral buckle.[6–10]

Despite the introduction of new diagnostic methods and treatment strategies for Coats' disease in recent years, few studies have been published to describe how these new advances have affected patients' anatomic and visual outcomes. Herein, we studied the population of Coats' patients who were diagnosed and treated at a tertiary care academic vitreoretinal practice over a 20-year period to compare treatment approaches and outcomes over 2 distinct 10-year periods.

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Methods

This was a retrospective review of all consecutive patients aged 18 years or younger at presentation with Coats' disease treated by multiple vitreoretinal specialists (E.G.B., B.W.M., G.J.J., E.A.P., T.H.M., C.A.T., L.V., P.M.) at the Duke University Eye Center between January 1, 1995, and September 30, 2015, and who were followed up for at least 12 months. The study was designed in accordance with the tenets of the Declaration of Helsinki, was approved by the Duke University Institutional Review Board, and complied with the Health Insurance Portability and Accountability Act. Patients with a diagnosis code of exudative retinopathy (International Classification of Diseases, Ninth Revision, code 362.12) were identified using the Duke Enterprise Data Unified Content Explorer database. Patients were excluded if they did not have clinical evidence of Coats' disease, were treated at another institution, or had missing or incomplete records.

Patients were diagnosed clinically with Coats' disease based on the following criteria: presence of idiopathic retinal telangiectasia with or without intraretinal or subretinal exudation and with or without exudative retinal detachments. Demographic information, visual acuity (VA) at several time points (initial presentation, the 12- to 18-month follow-up, the intermediate period for decade 1, and the final follow-up) when available were recorded, and details of each examination were recorded. Pediatric patients usually were examined and treated under anesthesia. Fundus photography and fluorescein angiography were performed using a RetCam 2 (Clarity Medical Systems, Pleasanton, CA), Zeiss FF450 (Carl Zeiss Meditec, Okerkochen, Germany), or Optos 200Tx (Optos, Inc., Dunfermline, United Kingdom) imaging device at the discretion of the treating physician.

Eyes then were divided into 2 groups based on year of presentation: decade 1 (1995−2005) and decade 2 (2006−2015). Furthermore, eyes were divided into 5 stages of disease at the time of presentation as previously described by Shields et al.[11] Briefly, stages 1, 2A, 2B, 3A, 3B, 4, and 5 are defined as follows: stage 1 corresponds to eyes with retinal telangiectasia only; stage 2A refers to eyes with retinal telangiectasia and extrafoveal exudation; stage 2B denotes eyes with retinal telangiectasia and foveal exudation; stage 3A represents eyes with retinal telangiectasia, exudation, and subtotal retinal detachment; stage 3B describes eyes with retinal telangiectasia, exudation, and total retinal detachment; stage 4 eyes demonstrate retinal telangiectasia, exudation, total retinal detachment, and secondary glaucoma; and stage 5 eyes are defined as blind eyes with retinal telangiectasia, exudation, total retinal detachment, and anterior chamber involvement or phthisis (advanced end-stage disease).

Management methods included observation (defined as no treatment for posterior manifestations of Coats' disease; some patients in this group still underwent cataract or strabismus surgery); ablative therapies including cryotherapy, 532-nm laser, or both; vitreoretinal surgery, which included any combination of subretinal fluid drainage, scleral buckle, pars plana vitrectomy, epiretinal membrane removal, and intraocular gas or silicone oil tamponade; IVT injection; or enucleation. After 2008, off-label intravitreal bevacizumab (IVB; 1.25 mg or 0.625 mg; Genentech, Inc., South San Francisco, CA) was given to selected eyes based on treating physician preference. Anatomic disease resolution was defined as resolution of exudates, exudative subretinal fluid, or both. When possible, best-corrected VA was obtained at each clinical visit. Visual acuity was measured using Snellen or Early Treatment Diabetic Retinopathy Study charts. In young children who could not recognize the letters or numbers on Snellen or Early Treatment Diabetic Retinopathy Study charts, VA was measured using Allen pictures or HOTV charts. Snellen VA equivalent of 20/125 was selected as a cutoff to determine eyes that maintained functional vision at the end of follow-up.

Statistical Analysis

Snellen VA equivalent was converted to logarithm of the minimum angle of resolution (logMAR) units for analysis. Snellen acuities for counting fingers, hand movements, light perception, and no light perception (NLP) measured at 2 feet were approximated as described previously.[12] Snellen acuities for measurements made at other distances were extrapolated from those obtained at 2 feet. Only 1 eye from each patient was included in statistical analysis. In patients with bilateral involvement, the worse eye was included in statistical analysis. For paired analysis (comparison of same eyes at different time points), only eyes that had documented Snellen VA equivalent at both of the visits being compared were included. The Mann−Whitney U test and Kruskal−Wallis test were used to compare continuous variables between groups and among groups, whereas the Fisher exact test was used to compare categorical variables between groups. The Wilcoxon signed-rank test was used to compare paired data. A P value of 0.05 or less was considered statistically significant. Statistical analysis was performed using SAS software version 9.3 (SAS Institute, Inc., Cary, NC).

Results

Baseline Characteristics of Study Participants

A total of 55 patients with complete medical records who were treated for Coats' disease at our institution were identified. A total of 16 patients were excluded: 14 had follow up less than 12 months and 2 were adults older than age 18 years at the time of presentation. Thirty-nine patients met inclusion criteria. Three patients demonstrated asymmetric bilateral disease at presentation (decade 1: stages 2B and 1 and stages 3B and 5; decade 2: stages 3A and 1). Only 1 eye from each patient was included in statistical analysis, and the better eye from patients with bilateral disease was excluded.

Baseline demographics and ocular characteristics of the final cohort of 39 eyes of 39 patients who met inclusion criteria are shown in Table 1 and are consistent with previously published reports.[1,13−19] There was no significant difference in baseline demographic characteristics between patients in the 2 decades. There was a trend for eyes in decade 1 to demonstrate more advanced stages of disease than those in decade 2: 47% of eyes in decade 1 demonstrated stage 3B to 5 disease (9 of 19 eyes) compared with 20% of eyes in decade 2 (4 of 20 eyes; $P = 0.1$).

Given the small number of eyes with advanced disease in decade 2, eyes from both decades were combined to examine the relationship between stage of disease and presenting VA. Eyes with more advanced stages of disease presented with worse initial VA ± standard deviation (SD; stage 2A, 0.12±0.13 logMAR [n = 4]; stage 2B, 0.87±0.48 logMAR [n = 5]; stage 3A, 1.78±1.03 logMAR [n = 15]; stage 3B, 2.95±0.21 logMAR [n = 7]; stage 4, 3.20 logMAR [n = 1]; stage 5, 3.20 logMAR [n = 1]; $P = 0.001$). Correspondingly, there was a trend for eyes in decade 1 to demonstrate worse VA than eyes in decade 2 ($P = 0.1$).