

Coats' disease: trends and long-term treatment outcomes in a tertiary referral centre

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Abstract

Background: The long-term treatment outcomes in Coats' disease – particularly in the era of newer pharmacotherapies such as anti-vascular endothelial growth factor (VEGF) agents and depot steroids – are poorly understood.

Aim: To describe the clinical features and treatment outcomes of 148 eyes with Coats' disease assessed in a referral centre over 30 years.

Materials and methods: We conducted a retrospective chart review of patients diagnosed with Coats' disease between 1 June 1987 and 31 July 2017. The demographic, clinical and treatment data were collected and long-term functional and anatomical outcomes were analysed based on the treatment either with conventional therapy (cryo/laser) or along with adjuvants like intravitreal steroids or anti-VEGFs.

Results: The mean age at presentation was 15.22 years (median 11). Familial exudative vitreoretinopathy was the most common referral diagnosis, 76/148 (51.5%), followed by Coats' disease, 37/148 (25%), and retinoblastoma, 35/148 (23.6%). Stage 3B was most common at presentation (31.8%), followed by 2B (22.3%) and 2A (16.9%). A total of 107 patients were treated either with conventional therapy or in combination with adjuvants. The mean follow-up period was 24.95 months. The visual acuity improved from baseline logMAR 2.17 (Snellen-20/2958) to logMAR 1.88 (Snellen-20/1517) at final follow-up ($p = 0.004$). The improvement in visual acuity was better when the presenting BCVA was <1 logMAR (Snellen 20/200), $p = 0.004$. No statistically significant change in BCVA was noted between conventional and adjuvant groups, $p = 0.5$. However, the final anatomical outcome was good in 78/99 (78.8%) in the conventional group and 45/49 (91.8%) in the adjuvant group, respectively ($p = 0.046$).

Conclusion: In this series of patients with Coats' disease over three decades, the use of intravitreal steroids or anti-VEGFs as adjuvants resulted in better anatomical outcomes. A better baseline visual acuity, lower stage of the disease, and older age at presentation were found to be the factors leading to favourable visual outcomes.

Summary

In the current series of 148 eyes with Coats' disease, adjuvant treatment with intravitreal steroids or anti-VEGFs resulted in better outcomes as compared with conventional cryotherapy or laser photocoagulation alone. Patients with Coats' disease who had presented with better visual acuity at baseline, lower stage of the disease and older age had better final visual outcomes.

Keywords: adjuvant treatment, anti-VEGF, Coats' disease, cryotherapy, exudation, laser treatment, laser treatment outcomes, retinal telangiectasia, stages, steroids

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Introduction

Coats' disease is an idiopathic, vascular disorder of the retina which presents with telangiectasia, intraretinal or subretinal exudation leading to exudative retinal detachment (RD). It chiefly presents unilaterally in young males without a family history of the disease.¹ Clinical spectrum of Coats' disease is broad, ranging from a mild asymptomatic retinal telangiectasia to exudative RD, phthisis or neovascular glaucoma in advanced cases. The peculiar clinical and imaging features help in differentiating Coats' disease from other life-threatening clinical entities like retinoblastoma as both can present with leucocoria.²

The demographic and clinical features of Coats' disease have been well described in the literature.^{1,3-5} Traditionally, the disease has been treated with conventional ablative therapies like cryotherapy (cryo) or laser photocoagulation. However, with the advent and widespread use of newer drugs like anti-vascular endothelial growth factor (VEGF) or depot steroids, the outcomes seem to have improved.⁶ The intraocular level of VEGFs and other inflammatory mediators are significantly increased in eyes with Coats' disease.⁶ Steroids seem to act by reducing the exudation, which is the hallmark of this disease, and the recent use of steroid implants in macular edema of varied aetiology is well described.⁷⁻¹¹ Previous studies have described treatment outcomes in Coats' disease based on traditional therapeutic modalities. Despite the widespread use of new treatment strategies, very few studies have taken into account the results of newer pharmacotherapy-based treatments. This study was undertaken to look into the clinical profile and long-term treatment outcomes including adjuvant therapy in cases of Coats' disease in a tertiary-care hospital.

Materials and methods

Subjects and study design

The Prasad Eye Institute Ethical Committee (LEC LVPEI Sep 2017-25) approved this retrospective study. The medical case records of 199 patients with the clinical diagnosis of Coats' disease, who presented to a tertiary referral centre from 1 June 1987 to July 2017, were retrieved from the electronic medical record using the search word Coats' disease. The study protocol adhered to the tenets of the Declaration of Helsinki. Written informed consent for inclusion had been obtained from all the patients.

Demographic and clinical profile. For the purpose of the study, demographic details such as age, gender, laterality, presenting complaints and diagnosis by referral ophthalmologists were recorded.

Data on comprehensive ophthalmic examination including best-corrected visual acuity (BCVA), slit lamp examination findings like neovascularisation of iris (NVI), anterior chamber cells, cataract, intraocular pressure (IOP) and retinal findings were collected. Fundus photography and fundus fluorescein angiography (FFA; Zeiss Visupac® FF4 and FF450-plus, Carl Zeiss, Dublin, CA), optical coherence tomography (OCT) using Zeiss Cirrus HD OCT and were analysed wherever available. Ophthalmic ultrasound B scan was also done in selected cases of media opacity to rule out tumours like retinoblastoma. Coats' retinopathy was diagnosed and classified as per Shields staging where stage 1 was retinal telangiectasia, stage 2A was telangiectasia with extra foveal exudation, 2B as foveal exudation, 3A was subtotal RD, 3B was total RD, stage 4 was RD with glaucoma and stage 5 was absolute eye. The stage of the disease, treatment modalities, treatment sessions, and follow-up visit details were recorded.

The patients who presented in the early years of the study period received the conventional treatment modalities and those who presented in later years had received the adjuvant treatments as per the clinical condition. The conventional treatment group comprises patients treated with cryo or laser alone, while those treated with adjuvant intravitreal steroid or anti-VEGF formed the adjuvant group. To analyse the data, patients were divided into subgroups based on the treatment received. The observation group constituted a confirmed case of Coats' retinopathy without active intervention, but a regular follow-up. Cryotherapy was done using triple freeze-thaw technique. Laser photoablation was done with double-frequency Nd YAG laser using laser indirect ophthalmoscope or slit lamp delivery. The laser was done directly to the telangiectasia and also to the adjacent capillary non-perfusion area where ever needed.

The intravitreal adjuvants used were triamcinolone acetonide (TA) 4 mg/0.1 mL, dexamethasone implant (Ozurdex, Allergan) and bevacizumab (1.25 mg/0.05 mL). The intravitreal injections were given under standard protocol under appropriate anaesthesia in a sterile

operating room. A combination of laser, cryo, intravitreal steroid or anti-VEGF was also used in some cases. Surgical interventions done were transscleral drainage of subretinal fluid, vitrectomy, enucleation, transscleral cyclophotocoagulation (TSCPC) or a combination of procedures.

The patients with incomplete records, follow-up of less than 1 year, partially treated elsewhere at presentation, were excluded from the study.

The patients were followed up at regular intervals. Visual acuity, IOP and fundus evaluation were done periodically.

Outcome measures. Visual acuity equal to 20/200 or better was defined as a good functional outcome.

A good anatomical outcome was defined as an intact non-phthisical, non-painful eye and poor anatomical outcome as a painful eye or phthisis bulbi. The Snellen visual acuity was converted to logMAR for statistical analysis.

Statistical analysis. The data were recorded in an Excel sheet (MS Office 2013) and analysed using SPSS version 18 for Windows (SPSS Inc., Chicago, IL, USA). Continuous variables were expressed as mean (median, range). Student *t* test was used to compare continuous variables and chi-square or Fisher's exact test for categorical variables.

Results

Demographic profile

Of 199 patients, 148 eyes of 148 patients who met study inclusion criteria were analysed. The mean follow-up period was 24.95 months. In the current retrospective study of 148 patients, 126 were males (85.1%) and 22 (14.9%) were females. The mean age at presentation was 15.22 years (range: 1–66) with a median of 11 years. The mean age at presentation of males was 15.13 years [95% confidence interval (CI): 12.66–17.61], and that of females was 15.73 years (95% CI: 8.68–22.78). A total of 37 (25%) patients were younger than 5 years of age. The right eye was involved in 85 (57.84%) and left eye in 63 (42.6%) patients.

Clinical profile

The most common presenting symptoms were reduced vision 96/148 (64.9%), leucocoria 31/148 (21%) and squint 25/148 (17%). A total

of 13 patients presented with a combination of decreased vision, squint and leucocoria.

The presenting referral diagnosis was retinoblastoma in 35 patients (23.6%), familial exudative vitreoretinopathy (FEVR) in 76 (51.5%) and Coats' disease in 37 (25%).

A total of 12 patients had other ocular or systemic diseases. Retinitis pigmentosa was noted in five eyes and seizure disorder was noted in two eyes. Of 148, 14 (9.5%) patients had neovascular glaucoma at presentation.

On clinical examination, the fundus findings noted were telangiectasia in 135 eyes (91.2%), intraretinal exudates in 134 eyes (90.5%), subretinal exudates in 130 eyes (87.8%), subfoveal exudate in 111 eyes (75%), exudative RD in 88 eyes (59.5%), vitreoretinal membranes in 34 eyes (23%) and vitreous haemorrhage in 14 eyes (9.5%). The stage of disease at presentation is given in Table 1.

FFA was available for 35 patients and capillary non-perfusion areas with telangiectasia were seen in 24 (88%), only telangiectasia in 4 (11%) and sclerosed vessels in 4 (11%) eyes. On analysis of OCT images (available for 16 patients), neurosensory detachment (NSD) was noted in 6 (37.5%), hyper-reflective subretinal deposits in 11 (68.75%), cystoid macular edema⁵ in 4 (25%) and foveal atrophy in 1 (6%).

Treatment

In this study of 148 patients, 107 (72.29%) received treatment and 41 (27.77%) patients were observed. In the treatment group, cryotherapy was done in 49 eyes (33.1%), laser in 64 eyes (43.2%), anti-VEGF in 29 eyes (19.6%), intravitreal steroids in 35 eyes (23.6%) and surgical intervention in 21 eyes (14.1%). The patients underwent surgical procedures like enucleation (4), subretinal fluid drainage (SRF) drainage (8), scleral buckle (3), Internal limiting membrane peeling (ILMP) (1), squint surgery (1), cataract extraction (2), TSCPC (2) and pars plana vitrectomy (2).

The total number of laser and cryo treatment sessions were significantly less with the use of adjuvant therapy. The total laser sessions were 1.43 in the conventional group and 0.73 in the adjuvant therapy group ($p = 0.001$). Similarly, mean cryo

Table 1. Demographic and clinical details.

Variable	Value	N
Total patients		148
Age	Mean (median)	15.22 years (11)
Sex		
Male		126 (85.1%)
Female		22 (14.9%)
Laterality	RE	85 (57.84%)
	LE	63 (42.6%)
Symptoms		
	Reduced vision	96/148 (64.9%)
	Leucocoria	31/148 (21%)
	Squint	25/148 (17%)
Referral diagnosis		
	Retinoblastoma	35 (23.6%)
	FEVR	76 (51.5%)
	Coats' disease	37 (25%)
Systemic association		
	Retinitis pigmentosa	5 (3.37%)
	Seizure disorder	2 (1.35%)
Follow-up	Mean	24.95 months
Clinical findings		
	Telangiectasia	135 (91.2%)
	Intra-retinal exudate	134 (90.5%)
	Subretinal exudate	130 (87.8%)
	Subfoveal exudate	111 (75%)
	Exudative RD	88 (59.5%)
	Vitreoretinal membranes	34 (23%)
	Vitreous haemorrhage	14 (9.5%)
FFA (n = 35)		
	Telangiectasia + Capillary nonperfusion	24 (88%)
	Only telangiectasia	4 (11%)

(continued)

Table 1. (continued)

Variable	Value	N
	Sclerosed vessel	4 (11%)
OCT (n = 16)		
	Neurosensory detachment	6 (37.5%)
	Hyper-reflective subretinal deposits	11 (68.75%)
	Cystoid macular edema	4 (25%)
	Foveal atrophy	1 (6%)
FEVR, familial exudative vitreoretinopathy; FFA, fundus fluorescein angiography; OCT, optical coherence tomography; RD, retinal detachment; RE, right eye; LE, left eye.		

sessions in the conventional group were 1.5, whereas it was 1.17 in the adjuvant group, though it was not statistically significant ($p = 0.20$). Mean number of steroid injections were 1.26, whereas mean number of anti-VEGF injections were 1.66 (range: 1–3 injections).

Visual and anatomical outcomes

The mean presenting BCVA was logMAR 2.17 ± 1.3 (SD) (Snellen 20/2958). A total of 14 eyes with no perception of light (PL) at presentation were excluded from visual acuity analysis. The BCVA at final follow-up was logMAR 1.88 ± 1.2 (SD) (Snellen 20/1517). The change in BCVA was statistically significant ($p = 0.004$, paired t test). The stage-wise baseline and final BCVA is given in Table 1. It was observed that the lower the stage of the disease, the better was the final BCVA. The visual outcome of better than 20/200 was noted in 40 eyes (27.02%) at baseline and in 42 eyes (28.37%) at the last follow-up.

In a subgroup analysis of the BCVA, mean and 95% CI at the presentation in patients with age less than 5 years and more than 5 years at the baseline was 2.66 (2.25–3.06) (Snellen 20/9141.76) and 1.97 (1.69–2.24) (Snellen 20/1866.50), respectively, and that on final follow-up was 2.25 (1.78–2.72) (Snellen 20/3556.55) and 1.75 (1.48–2.01) (Snellen 20/1124.68), respectively. The change in BCVA was statistically significant in both the subgroups ($p < 0.05$). The older the age of presentation, the better was the baseline visual acuity.

In a subgroup analysis of patients with BCVA of less than 1 logMAR and BCVA of more than 1 logMAR, it was found that the change in BCVA was significantly more in the group with BCVA less than 1 logMAR ($p = 0.004$). The initial and final BCVA in group with BCVA < 1 logMAR was 0.57 SD, 0.31 (0.47–0.67) and 0.53 SD, 0.37 (0.42–0.65), respectively, $p = 0.004$. The initial and final BCVA in group with BCVA > 1 logMAR was 2.85 SD, 0.91 (2.66–3.04) and 2.57 SD, 1.02 (2.35–2.80), respectively ($p = 0.348$). The visual improvement was noted to be better when the presenting BCVA was < 1 logMAR (Snellen 20/200). The change in BCVA in the conventional treatment group (logMAR 1.87 ± 1.34) and that in the adjuvant group (logMAR 1.9 ± 1.2) was not statistically significant ($p = 0.5$).

The final anatomical outcome was good in 83.1% (123/148), absolute eye in 13.5% (20/148) and phthisis in 3.3% (5/148) eyes. Furthermore, the subgroup analysis of the observation group and treatment group was done. In the observation group, where the patients were not given any active treatment, however they were followed up regularly, the anatomical outcome was good in 58.53% (24/41), absolute eye in 34.14% (14/41) and phthisis in 7.31% (3/41) eyes, respectively. In the treatment group, the anatomical outcome was good, absolute eye or phthisis in 92.52% (99/107), 5.6% (6/107) and 1.8% (2/107) eyes, respectively. It was evident that the active intervention resulted in better anatomical outcome. The final anatomical outcome was good in 78/99 (78.8%) and 45/49 (91.8%) in the conventional and adjuvant groups, respectively ($p = 0.046$) (Table 2).

The stage of the disease, at the final follow-up, is as given in Table 1. The stage of disease was stable in 58.1% (86/148), worsened in 18.2% (27/148) and regressed in 23.6% (35/148) at the final follow-up. The disease progressed, regressed and remained stable in 20.2% (20/99), 20.2% (20/99), and 59.59% (59/99) and 14% (7/49), 30.61% (15/49), and 55.10% (27/49) in conventional and adjuvant group, respectively.

Discussion

This study is a large retrospective series on demographic and clinical profile of patients with Coats' disease with a long-term follow-up. Even though Coats' disease can occur at any age, it was found to be more common in first and second decade of

life. The mean age of patients in this study was 15.3 years which is comparable with the study by Rishi *et al.* in Southern India (mean age: 15.67 ± 11.6 years), but is more than by Shields *et al.* (mean age: 11 years).^{1,3} A male preponderance (85.1%) as reported earlier in other studies was noted in this study also.^{1,3,4,12} In this series, we found that children less than 5 years presented with severe disease with profound vision loss when compared with older children which is similar to the earlier reports by Daruich and Shields *et al.*¹³ FEVR was the most common referral diagnosis in this group unlike that from Shield's group where it was retinoblastoma.¹ The findings of the study drive an important message for the general ophthalmologist to promptly refer such patients. Most of the patients presented with reduced vision as the presenting symptom as reported in other studies.^{3,4,14} Coats' disease is characterised by retinal telangiectasia and intra- or subretinal exudation and these were the common findings in this study also. At presentation, stage 3B disease was noted in 47% of eyes, as reported by Rishi *et al.* but unlike that reported by Al-Qahtani *et al.* (stages 3A and 2B) and B Morris *et al.* (stages 2B and 3A).^{4,14} At presentation, 9.5% had neovascular glaucoma, while it was 8% in Shield's group.¹ The common FFA features were telangiectasia and capillary non-perfusion areas in this series also as reported earlier.⁴

OCT is a useful tool to diagnose the type of macular involvement and to document the extent and type of macular involvement on follow-up. In addition, it helps to tailor the need for intravitreal treatment and prognosticate the visual recovery. In this study, subretinal exudates and NSD were most common OCT findings, even though the scans were available only for 10.81% patients. The presence of subretinal fibrosis at fovea is a predictor of poor visual outcome and an OCT-based classification has been proposed.¹⁵

The natural history of Coats' disease is varied. A study on natural history showed the disease progression in 50% of patients with time.¹⁶ Cases of spontaneous reattachment of exudative detachment were also reported.^{17,18} The treatment for Coats' disease has evolved with time. The aim of treatment is to control the disease progression by closing the abnormal leaky telangiectatic retinal vessels. Reduced leakage and exudation facilitate reattachment of retina and globe salvage by preventing complications like neovascular glaucoma and ultimately phthisis bulbi. The standard

Table 2. Anatomical and functional outcomes.

Stage of disease	Number of eyes at baseline N (%)	Pre BCVA Mean (SD)	Number of eyes received treatment/ observed	Mode of treatment (N)	Final BCVA Mean (SD)	Number of eyes at final follow-up N (%)	Number of eyes with final BCVA >20/200	Anatomical outcome (nos) (N)
1	5 (3.4)	1.04 (1.381)	5/0	Cryo 3 Laser 3 Steroid 4 Anti-VEGF 0	0.88 (0.756)	16 (10.8)	50% (2/4)	Good – 4 Absolute eye – 0 Phthisis bulbi – 1
2A	25 (16.9)	1.54 (1.207)	19/6	Cryo 9 Laser 12 Steroid 10 Anti-VEGF 4	1.44 (1.15)	48 (32.4)	26.08% (6/23)	Good – 22 Absolute eye – 3 Phthisis bulbi – 0
2B	33 (22.3)	1.98 (1.17)	26/7	Cryo 9 Laser 17 Steroid 4 Anti-VEGF 9	1.55 (1.09)	14 (9.5)	23.33% (7/30)	Good – 30 Absolute eye – 2 Phthisis bulbi – 1
3A	19 (12.2)	1.99 (1.35)	11/8	Cryo 5 Laser 7 Steroid 7 Anti-VEGF 7	1.18 (.92)	12 (8.1)	47.05% (8/17)	Good – 16 Absolute eye – 3 Phthisis bulbi – 0
3B	47 (31.8)	2.55 (1.2)	31/16	Cryo 13 Laser 20 Steroid 2 Anti-VEGF 4	2.57 (1.2)	29 (19.5)	34.28% (12/35)	Good – 34 Absolute eye – 10 Phthisis bulbi – 3
4	4 (2.7)	2.23 (1.5)	3/1	Cryo 3 Laser 1 Steroid 0 Anti-VEGF 0	1.2 (.42)	3 (2)	0% (0/3)	Good – 4 Absolute eye – 0 Phthisis bulbi – 0
5	15 (10.1)	3.5 (.53)	12/3	Cryo 7 Laser 4 Steroid 8 Anti-VEGF 5	3.67 (.51)	26 (17.6)	0% (0/12)	Good – 13 Absolute eye – 2 Phthisis bulbi – 0

BCVA, best-corrected visual acuity; SD, standard deviation; VEGF, vascular endothelial growth factor.

conventional therapies used for Coats' disease are cryotherapy and laser photocoagulation, to target the leaky abnormal vessels.¹⁹ Cryotherapy was preferred in advanced cases in one of the earlier reports and 42% underwent cryotherapy in the series reported by Shield's *et al.*^{20,21} The cryotherapy could act even in the presence of shallow sub-retinal fluid where laser photocoagulation would have limited utility. A total of 42% of patients in our series received laser photocoagulation as the primary treatment which was comparable with recent studies (Rishi *et al.* – 33%, Al-Qahtani *et al.* – 64%, A Mulvihill *et al.* – 92%), unlike the earlier studies (Shields *et al.* – 13%).^{3,4,22}

The pathogenesis in Coats' disease is the abnormal permeability of vessels due to the breakdown of blood retinal barrier leading to excessive exudation. Steroids and anti-VEGF agents seem to

have a therapeutic role by reducing exudation and restoring the blood retinal barrier.^{23,24} Increasing levels of intra-ocular VEGFs were found with increasing severity of Coats' disease.^{6,25,26} The increasing evidence on the adjuvant role of anti-VEGFs to reduce the exudation in Coats' disease has been reported by various authors.^{19,27–32} Ramasubramanian *et al.* cautioned about the use of anti-VEGF agents as they noticed vitreo retinal fibrosis, while that by Ray *et al.* reported good outcomes with the use of anti-VEGF agents.^{31,33} There are few reports on the use of intravitreal steroids in the resolution of exudative RD and subfoveal exudates in Coats' disease.^{34,35} Bergstrom and Hubbard³⁶ had cautioned about the use of intravitreal TA along with cryo in exudative RD as they noticed delayed complications like rhegmatogenous RD with proliferative vitreo-retinopathy in three out of five patients in their

series. Intravitreal injections (steroids or anti-VEGF) were given in 39.9% of eyes in this study. The disease had either stabilised or regressed in 85% of those who received intravitreal treatment and 91% had good anatomical outcomes. A good anatomical outcome was noted in 92% of patients who underwent treatment compared with 58% in the observation group. The data presented here are a retrospective chart review of the cases which signifies the physician discretion to offer treatment as per the clinical condition in actual clinic setting.

Consistent with our findings, earlier reports have demonstrated that lower the stage of the disease at presentation, better was the BCVA at final follow-up.^{13,37} Rishi *et al.*³ showed that there was no significant change in visual acuity at presentation and at final follow-up. In our series, BCVA at last follow-up was significantly improved compared with baseline. This difference could be possibly explained using intravitreal agents like intra-vitreal steroids and anti-VEGFs to target the exudation. Li *et al.*²⁹ had reported improvement in visual acuity in 41.8% of stage 3B Coats' disease treated with a combination of intravitreal ranibizumab along with cryo and laser. The final visual acuity was worse than 20/200 in 71% of patients in this series which is less than that reported by Rishi *et al.* (89.9%) but more than that reported by Al-Qahtani *et al.* (59%).^{3,4} The improvement in visual acuity was statistically significant if presenting visual acuity was better than logMAR 1 (Snellen 20/200) ($p < 0.004$). Meanwhile the improvement in BCVA in the group who received adjuvant therapy was better than in conventional group, but it was not statistically significant. This was noted in the other studies also.³⁴

This being a retrospective study, the authors admit the inherent weakness of selection bias of treatment groups and non-availability of investigations like OCT and FFA for all the patients, but Coats' disease is a rare disease and this article presents a real-life data on presentation, treatment and clinical outcomes of Coats' disease.

In contrast to conventional treatment, better anatomical outcomes were noted with the use of adjuvant intravitreal anti-VEGFs or steroids, which help in faster resolution of exudates and facilitate more precise delivery of laser or cryotherapy. The findings of this article highlight the fact that pharmacotherapeutic adjuvants have a

role to play in reducing treatment sessions and should be incorporated in current treatment algorithm.

In conclusion, we present a series of 148 eyes with Coats' disease treated with conventional and adjuvant therapy. The factors favouring better visual outcomes were lower stage of the disease, better presenting BCVA and older age at presentation. The use of either intravitreal steroids or anti-VEGF leads to better anatomical outcomes but equivocal functional outcomes.

Author contributions

Data collection and analysis, initial draft of the manuscript: AO. Data analysis, manuscript writing, supervision of conduct of study: DB. Final draft of manuscript, supervision of the study: SJ

Conflict of interest statement

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