External Drainage Alone Versus External Drainage With Vitrectomy in Advanced Coats Disease

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• PURPOSE: To evaluate the efficacy of transscleral drainage of subretinal fluid (TDSRF) with and without pars plana vitrectomy (PPV), which are performed as an adjunct to ablative therapies in advanced Coats disease.

• DESIGN: Retrospective, comparative case series. Patient Population: A total of 31 eyes from 31 consecutive patients with advanced Coats disease at an average age of 47.8 ± 34.2 (2-156) months at the time of the surgery, who underwent TDSRF alone or in combination with PPV.

• METHODS: Main outcome measurements were LogMAR visual acuity values, anterior and posterior segment findings, need for further surgery, laser photocoagulation, and anti-vascular endothelial growth factor (anti-VEGF) treatment. Anatomical success was defined as the maintenance of retinal reattachment without any further surgery during follow-up.

• RESULTS: Sixteen patients underwent TDSRF alone, and 15 patients underwent combined TDSRF and PPV. Mean follow-up time was 34.8 ± 32.9 months (6-128). Anatomical success rate of combined TDSRF and PPV was higher than that of TDSRF alone (93.8% vs 75%, respectively). The incidence of epiretinal membrane formation, number of laser photocoagulation procedures, and anti-VEGF treatments were statistically significantly higher in the group that underwent TDSRF alone than in those who had combined TDSRF and PPV in the long term. There was an improvement in vision in only 4 eyes (12.9%) (all >3 years old at presentation) during the follow-up.

• CONCLUSIONS: Combined TDSRF and PPV appears to be more effective in controlling the disease than TDSRF alone, as an adjunct to ablative procedures for the treatment of advanced Coats disease. The need for additional treatment is much less with the addition of PPV. (Am J Ophthalmol 2021;222:6–14. © 2021 Elsevier Inc. All rights reserved.)

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COATS DISEASE, FIRST DESCRIBED IN 1908, IS commonly a unilateral sporadic disease occurring mostly in young men. It is characterized by retinal vascular anomalies leading to intraretinal and subretinal lipid exudation and, later, exudative retinal detachment (ERD).[1] Shields and associates[2] divided Coats disease into the following 5 stages as follows: Stage 1, presence of retinal telangiectasia; Stage 2, telangiectasia and exudation (Stage 2A, extrafoveal exudation; Stage 2B, foveal exudation); Stage 3, ERD (Stage 3A, subtotal detachment; Stage 3B, total detachment); Stage 4, total RD and neovascular glaucoma; and Stage 5, advanced end-stage disease. Ablation therapy aimed at obliterating abnormal telangiectatic vessels by using laser photocoagulation or cryotherapy has been the mainstay of treatment for Coats disease for decades.[3] Furthermore, following a marked increase in the intravitreal vascular endothelial growth factor (VEGF) concentration in patients with Coats disease, anti-VEGF agents are commonly used for adjuvant treatment, together with conventional ablative therapies.[4–6]

Previous studies have shown favorable visual and anatomical outcomes in patients with early stages of Coats disease who were treated with anti-VEGF therapy alone or in combination with ablative therapies.[7,8] However, some researchers have emphasized that cryotherapy and anti-VEGF therapy may contribute to the development of an epiretinal membrane (ERM), preretinal fibrosis, and even tractional retinal detachment (TRD).[9,10] In advanced Coats disease, the visual prognosis is poor due to photoreceptor cell injury secondary to massive submacular exudation and dysfunction of retinal pigment epithelium.[11] Furthermore, cryotherapy and laser photocoagulation cannot be performed effectively because of the massive subretinal exudation and ERD. Laser therapy requires the retina to be in contact with the retinal pigment epithelium to produce effective burns and coagulation. Therefore, it is mainly effective to obliterate abnormal vessels located on the attached retina. Although cryotherapy has an ablative effect on telangiectatic vessels located on the detached retina, massive subretinal exudation and total ERD can obscure visualization of abnormal peripheral vessels in patients with advanced Coats disease. Accordingly, many surgeons prefer to drain the subretinal fluid (SRF) and exudate using a transscleral method for effective ablation of abnormal vessels during treatment of advanced Coats disease.[12–14] Pars plana vitrectomy (PPV) should be

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TABLE 1. Baseline Demographical and Clinical Characteristics of Patients with Advanced Coats Disease

	TDSRF	TDSRF + PPV	Total	*P*
Patients	16 (51.6)	15 (48.4)	31	
Mean ± SD age at surgery, mo	40.4 ± 37.4	51.2 ± 28.2	45.6 ± 33.1	.187[a]
Mean ± SD follow-up time, mo	35.3 ± 34.9	34.1 ± 31.5	34.8 ± 32.9	.924[a]
Females	2 (12.5)	4 (26.7)	6 (19.3)	.318[c]
"Fix and follow" or better vision	3 (18.7)	4 (26.7)	7 (22.6)	.598[c]
Epiretinal membrane	–	2	2	
Main first sign and symptom				
Leukocoria	8 (50)	7 (46.7)	15 (48.4)	.853[b]
Strabismus	6 (37.5)	7 (46.7)	13 (41.9)	.605[b]
Vision loss	2 (12.5)	1 (6.6)	3 (9.7)	.583[c]
Stage of Coats disease				
3A	6 (37.5)	5 (33.3)	11	
3B	8 (50)	9 (60.0)	17	
4	2 (12.5)	1 (6.6)	3	
5	–	–	–	

PPV = pars plana vitrectomy; TDSRF = transscleral drainage of subretinal fluid.
Values are n (%) or mean ± SD.
[a]Mann-Whitney *U* test.
[b]χ^2 test.
[c]Fisher exact test.

considered in cases of vitreoretinal traction and vitreoretinal interface problems.[15] Furthermore, a large amount of VEGF molecules can be removed from the vitreous through PPV. PPV accompanied by membrane peeling removes existing tractional components on the retina and may have a prophylactic effect against the possible progression of vitreoretinal traction secondary to ablative therapies, such as anti-VEGF and cryoapplication. However, PPV carries the risk of intraoperative complications such as iatrogenic retinal breaks.[15]

Present knowledge about the effect of transscleral drainage of subretinal fluid (TDSRF) on the clinical course of advanced Coats disease, either alone or in combination with PPV, remains limited. To the best of the present authors' knowledge, no study has evaluated the role of PPV in addition to external drainage in advanced Coats disease.

This study reviewed the outcomes of patients with advanced Coats disease treated with ablative therapies, accompanied by TDSRF alone or in combination with PPV, and the differences in outcomes according to early versus late presentation.

MATERIALS AND METHODS

THIS RETROSPECTIVE CASE SERIES STUDY FOLLOWED THE tenets of the Declaration of Helsinki and was approved by the Gazi University Research Ethics Committee. Written informed consent was obtained from the parents of each participant of this study.

• **PATIENT POPULATION:** The medical records of patients with advanced Coats disease (Stage 3 or higher) treated with ablative therapies accompanied with TDSRF alone or in combination with PPV, between January 2009 and January 2019, were retrospectively analyzed. Patients with a history of any previous ocular intervention or those with less than 6 months of follow-up were excluded from the study. All surgeries were performed by 1 vitreoretinal surgeon (Ş.Ö.). In this historically controlled case series, the initial patients mostly underwent TDSRF alone, whereas subsequent patients mostly underwent TDSRF combined with PPV, which was determined by the surgeon's experience with previous surgeries. Patients were divided into the following 2 groups according to the type of adjunctive surgery: TDSRF alone or TDSRF combined with PPV.

• **SURGICAL TECHNIQUE:** All TDSRF surgeries were performed with the patients under general anesthesia. Briefly, the general surgical steps for TDSRF were 1) a scleral incision was performed in the area corresponding to the most massive SRF and exudate. 2) SRF and exudate were drained using a cotton bud. 3) Ocular tonus was maintained by using a valved 25-gauge trocar placed at the pars plana (if the retina was not located behind the lens, in order to enable safe entry through the pars plana) or by using a limbal incision (if the detached retina is located behind

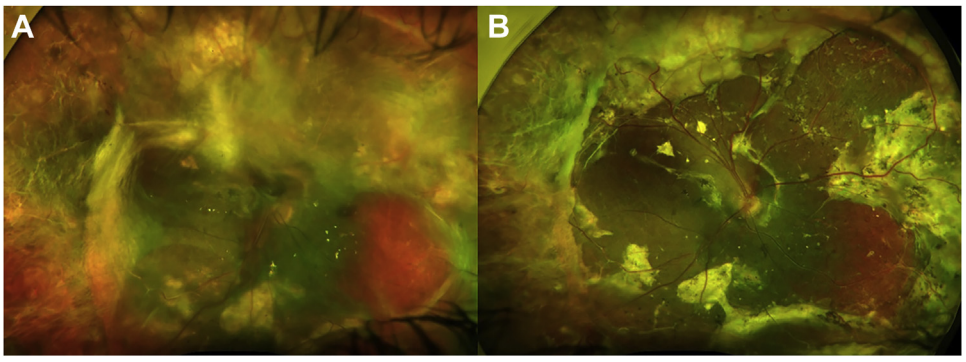

FIGURE 1. This patient was referred to the authors' hospital with a diagnosis of Stage 3B Coats disease of the right eye and underwent TDSRF when he was 2 years old. After that, he had multiple laser photocoagulation and cryotherapy sessions within 4 years of the follow-up. (A) Dense epiretinal membranes are causing tractional retinal detachment 4 years after initial TDSRF surgery. Vision was questionable light perception. He underwent PPV as a second surgery at 6 years old. (B) Completely attached posterior retina together with widespread peripheral subretinal fibrosis 1 year after PPV. The final visual acuity was limited to hand motion. PPV = pars plana vitrectomy; TDSRF = transscleral drainage of subretinal fluid.

the lens), as applicable. 4) A 25-guage trocar was placed at the pars plana in patients in whom it had not been placed previously when the detached retina behind the lens moved posteriorly after drainage of SRF. 5) Direct laser photocoagulation of abnormal dilated telangiectatic vessels was performed with a lighted endolaser probe through the trocar. 6) Cryotherapy was applied in areas of peripheral telangiectasia and adjacent avascularity under indirect ophthalmoscopy (not more than 2 quadrants); this step can also be performed using an operating microscope with indirect viewing systems (EIBOS; Haag-Streit, Mason, Ohio, USA) by placing a chandelier light on the pre-existing trocar. 7) Bevacizumab was injected intravitreally (1.25 mg/0.05 mL; Avastin) was administered at the end of the surgery.

In patients undergoing combined TDSRF and PPV, after the first 3 steps, three 25-gauge trocars were inserted through the pars plana, and a central core lens-sparing vitrectomy with membrane peeling was performed. PVD was attempted in the posterior pole until just anterior to arcuate vessels if possible and a complete separation of PVD until periphery was not aimed at all. Following that, the last 3 steps were performed with the addition of an air-fluid exchange before the intravitreal bevacizumab injection.

During the postoperative period, patients with recurrent exudation or leaky abnormal telangiectatic vessels were treated with laser photocoagulation under general anesthesia. Intravitreal bevacizumab injection was administered simultaneously if there was significant concomitant SRF.

• **MAIN OUTCOMES:** At baseline and during each postoperative visit, ophthalmologic examination findings were recorded, including visual acuity, anterior and posterior segment findings, and complications. The need for further ablative therapy, surgery, and anti-VEGF injection was also recorded. Anatomical success was defined as maintenance of retinal reattachment without any further surgery during follow-up. Functional success was defined as ambulatory or better vision (logMAR ≤1.7) at the last follow-up.

• **STATISTICAL ANALYSIS:** SPSS version 22.0 software (SPSS, Chicago, Illinois, USA) was used to analyze the collected data. The Shapiro-Wilk test was performed to determine whether the continuous variables were parametric or nonparametric. Mann-Whitney *U*, χ^2, and Fisher exact tests were performed to compare the clinical characteristics and surgical outcomes between the 2 groups, where appropriate. A *P* value ≤ .05 was considered statistically significant.

RESULTS

A TOTAL OF 31 EYES OF 31 PATIENTS WITH ADVANCED Coats disease were included in the study. The ablative therapies were accompanied by TDSRF only in 16 patients and by a combination of TDSRF and PPV in 15 patients. The mean age of patients at surgery was 45.6 ± 33.1 (range, 2-156) months. The average follow-up time was 34.8 ± 32.9 (range, 6-128) months. The mean follow-up time was 35.3 ± 34.9 months for the TDSRF-only group and 34.1 ± 31.5 months for the combined TDSRF and PPV group. At baseline, 7 (43.8%) of 16 cooperative patients had at least a "fix-and-follow vision" with a mean visual acuity of 2.28 ± 0.48 logMAR in the affected eye. Leukocoria (48.4%) was the most common initial sign, particularly in patients younger than 3 years of age (73.3% in 11 of 15 younger patients). Most patients had Stage 3 Coats disease. Table 1 shows the detailed baseline demographic and clinical characteristics of the patients.

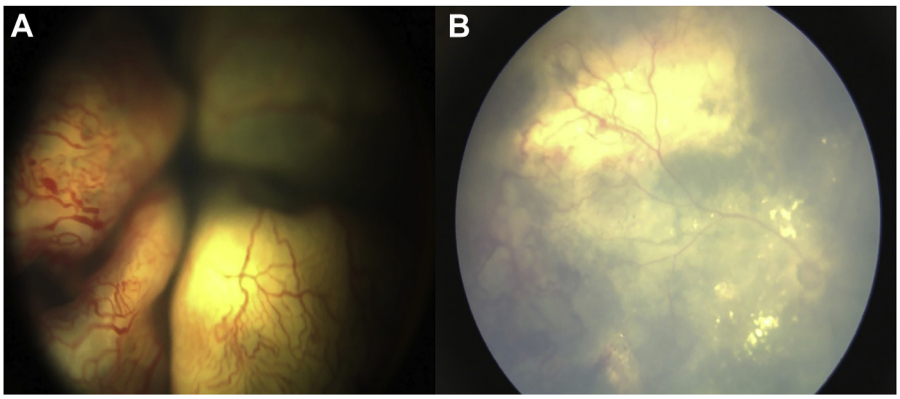

FIGURE 2. (A) Preoperative view of 2-year-old girl with stage 3B Coats disease shows total exudative retinal detachment and widespread abnormal telangiectatic vessels. This patient underwent combined TDSRF and PPV surgery. (B) Retina was reattached completely 1 month after the surgery. PPV = pars plana vitrectomy; TDSRF = transscleral drainage of subretinal fluid.

When the patients were divided into 2 groups according to the age of onset of disease, the ratio of eyes with Stage 3B (more advanced disease) was significantly higher in the patients younger than 3 years than in those older than 3 years. (73.3% vs. 37.5%, respectively; $P = .045$). However, the need for further surgery (13.3% vs. 12.5%, respectively), laser photocoagulation (33.3% vs. 31.3%, respectively), and anti-VEGF therapy (31.3% vs. 20%, respectively) were similar between the 2 groups. There were no significant differences in anatomical outcomes after the surgery between 2 groups.

In the TDSRF-alone group, 4 eyes (25%) required PPV as a second surgery within a mean follow-up time of 25 months for TRD (Figure 1). Therefore, anatomical success could be achieved in 12 eyes (75%) after the first surgery. In the combined TDSRF and PPV group, separation of the posterior hyaloid membrane could be achieved during vitrectomy in 13 eyes (86.7%), and retinal reattachment was achieved in all but 1 eye (93.8%) within a mean follow-up time of 34.1 ± 31.5 months (Figure 2). Differences in the anatomical success rates of the groups were not statistically significant ($P = .196$) (Table 2). The failed eye had Stage 4 Coats disease with neovascular glaucoma preoperatively and then developed hypotonia and seclusio pupillae after the surgery. Considering that the patient in whom surgery failed would not benefit from further surgery, intravitreal anti-VEGF injections were administered to stabilize the eye in order to prevent a painful eye and evisceration. Consequently, a quiet and nondeformed globe could be preserved. However, the retina remained detached and nonfunctional.

Ten patients required additional laser treatment, and 8 required anti-VEGF injection starting from a mean of 11 months after the initial surgery (Figure 3). The number of laser and anti-VEGF treatments required was significantly higher in the TDSRF-only group than in the combined TDSRF and PPV group at the 24- and 36-month follow-ups (Table 2).

Regarding the functional outcomes, 8 of 17 patients (47.1%) at cooperative ages had considerable vision (perception of hand motion or better) with a mean visual acuity of 1.91 ± 0.59 logMAR at the final visit. Ambulatory vision was achieved in only 2 eyes (6.4%), 1 of which had a vision of logMAR 1 (20/200), and the other had a vision of logMAR 1.3 (20/400). The remaining eyes had vision of light perception. Among the 17 patients at cooperative age, 4 patients (12.9%), all older than 3 years, had increased vision (Table 2); however, 2 patients (6.4%) experienced reduced vision, and the remaining patients had stable vision. None of the patients younger than 3 years of age had an improvement in vision.

Subretinal fibrosis (100%) was the complication most commonly observed in both groups (Figure 4). Subfoveal nodules were observed in 27 patients (87.1%) (Figure 5). Fundoscopic examination revealed TRD and marked ERM (Figure 1) in 4 and 6 patients, respectively, in the TDSRF-only group, whereas neither ERM nor TRD were observed in the combined TDSRF and PPV group, and the differences in ERM formation were statistically significant ($P = .011$) (Table 3). Yellow crystals in the anterior chamber, considered a bad prognostic sign, together with posterior synechia and hypotonia, were also noted in an eye with Stage 4 disease in the combined group (as mentioned above). None of the patients progressed to more advanced Coats disease, and none of the eyes required removal because of a painful red eye or deformed globe.

DISCUSSION

LESS INVASIVE TREATMENTS SUCH AS CRYOTHERAPY, laser photocoagulation, and anti-VEGFs have been reported to be effective when used alone or in combination for the treatment of early stages of Coats disease. However, advanced Coats disease requires more than these

TABLE 2. Results in the Treatment Groups

	TDSRF (n = 16)	TDSRF + PPV (n = 15)	*P*
Eyes with anatomical success after initial surgery	12 (75)	14 (93.3)	.186[a]
Eyes requiring further surgery	4 (25)	–	.058[a]
Eyes requiring further laser therapy	7 (43.7)	3 (20)	.152[a]
Eyes requiring additional anti-VEGF injection	5 (31.2)	3 (20)	.382[a]
Eyes showing an increase in vision during follow-up	1 (6.3)	3 (20)	.196[a]
Mean number of the laser therapies applied per patient at each time point (of the patients receiving laser therapy)			
At the end of 12th mo	0.53 (26.7)	0.29 (14.3)	.168[a]
At the end of 24th mo	1.08 (41.7)	0.58 (25)	**.044**[b]
At the end of 36th mo	1.5 (70)	1.0 (33.3)	**.036**[b]
Mean number of the anti-VEGF therapies applied per patient at each time point (% of the patients receiving anti-VEGF therapy)			
At the end of 12th mo	0.47 (26.7)	0.14 (14.3)	.070[a]
At the end of 24th mo	0.83 (41.7)	0.33 (16.7)	**.045**[b]
At the end of 36th mo	1.3 (50)	0.55 (33.3)	.087[b]
Mean number of total additional procedures applied per patient at each time point (% of the patients receiving any of additional procedure)			
At the end of 12th mo	1 (26.7)	0.43 (14.3)	.070[a]
At the end of 24th mo	2.08 (58.3)	0.92 (25)	**.027**[b]
At the end of 36th mo	3.2 (90)	1.55 (33.3)	**.021**[b]
Number of the patients still under observation at each time point			
At the end of the 12-mo follow-up	15	14	
At the end of the 24-mo follow-up	12	12	
At the end of the 36-mo follow-up	10	9	

Anti-VEGF = anti-vascular endothelial growth factor; PPV = pars plana vitrectomy; TDSRF = transscleral drainage of subretinal fluid.

Values are n (%). Total additional procedures involved further surgery, laser photocoagulation, and anti-VEGF therapies. Bold values indicate the statistically significant values.

[a]Fisher exact test.

[b]χ^2 test.

treatments, as expected.[16] This is because ablative therapies such as laser and cryotherapy cannot be performed effectively owing to massive subretinal exudation in advanced disease. To date, external drainage of SRF, PPV, and a combination of these procedures have been performed to increase the effectiveness of ablative therapy.[15] At this point, there is no consensus on which surgery should be performed, and surgical procedure is determined based on the surgeon's preference.

Previous studies evaluating the efficacy of TDSRF without PPV in the treatment of advanced Coats disease demonstrated favorable outcomes after the ablative therapy with TDSRF alone.[13,17] Stanga and associates[13] performed a combination of TDSRF, anti-VEGF, and laser photocoagulation therapy in 8 patients with advanced Coats disease and followed the patients for an average of 33 months; retinal reattachment was maintained in all patients during the follow-up.[13] Similarly, Cai and associates[17] treated 25 patients with advanced Coats disease using a combination of TDSRF, direct laser photocoagulation, and anti-VEGF therapy. They reported that retinal attachment was maintained in 96% of patients in a short-term follow-up (~10 months).[17] However, Li and associates[18] combined ablative therapy with PPV and scleral buckling or TDSRF in the patients and reported that 4 patients (50%) who were not initially treated with PPV required further PPV due to progressive TRD at a mean onset of 20 months after the initial treatment.[18] In contrast, only 1 patient undergoing early PPV combined with ablative therapy required further PPV surgery due to ERM in their studies. They emphasized that prophylactic vitrectomy might protect against the development of subsequent TRD.[18] Furthermore, Bhat and associates[12] performed a combination of TDSRF, cryotherapy, and bevacizumab and reported that 3 patients (42.9%) developed TRD after the surgery. Accordingly, in the present study, TRD developed in 25% of the patients in the nonvitrectomized group. Given the outcomes, TDSRF seems to be a relatively effective procedure as an adjunct to ablative treatments to achieve retinal reattachment in the short term. However, this positive outcome may be lost in the long term because of the potential risk for subsequent TRD, which may develop as a part of the disease itself or

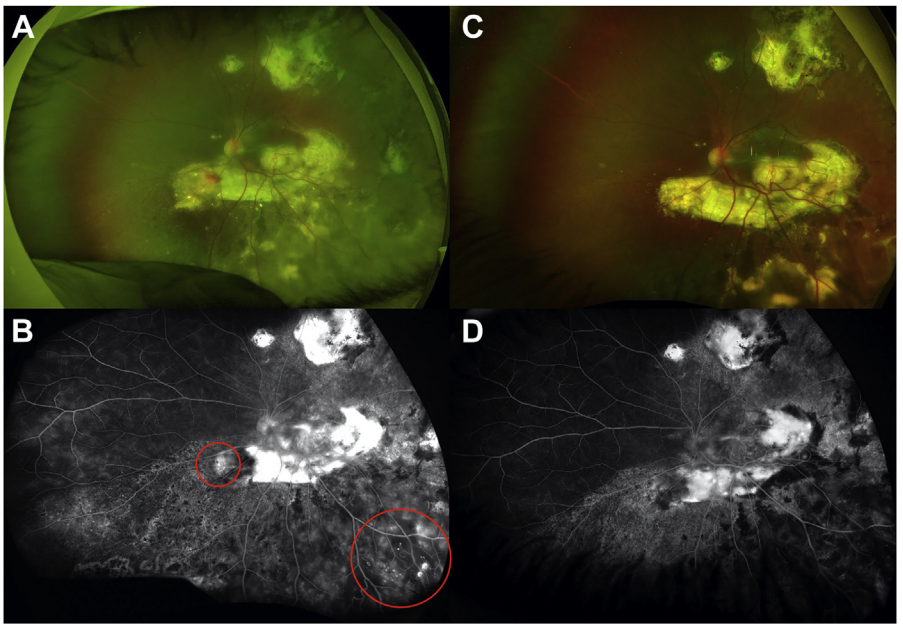

FIGURE 3. This patient with Stage 3B Coats disease underwent a combined TDSRF and PPV procedure at 5 years old. (A) View of the fundus 2 years after the surgery shows a subfoveal nodule and subretinal fibrosis around the inferior arcuate vessels and superior to the macula, and SRF inferiorly. Note that there is no epiretinal membrane formation. The vision was counting fingers at 2 m. (B) Late phase of the fundus fluorescein angiography of the same patient. Note the leaky abnormal vessels at the inferotemporal and inferonasal retina (red circles) corresponding to the area of SRF (demonstrated in A), indicating the need for additional treatment. (C) SRF has completely resolved, leaving subretinal exudates around the vessels 6 months after the combined laser photocoagulation and anti-VEGF therapies. (D) Effective ablation of the leaky vessels at inferior fundus with laser photocoagulation. The final vision remained unchanged. PPV = pars plana vitrectomy; SRF = subretinal fluid; TDSRF = transscleral drainage of subretinal fluid.

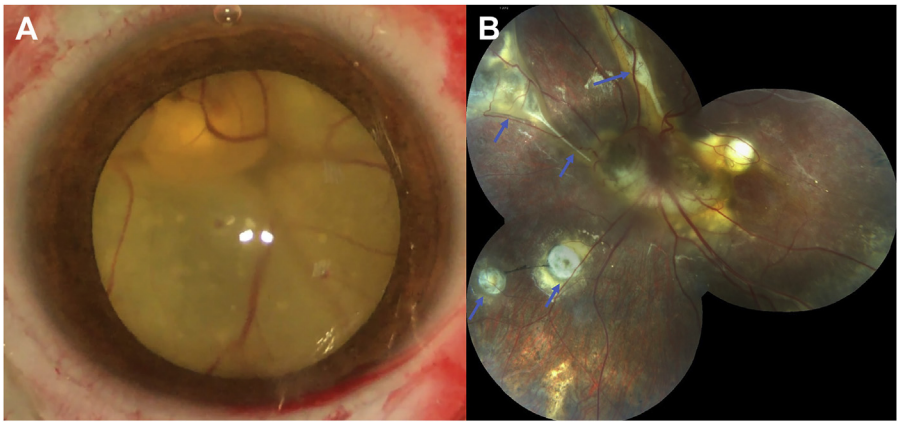

FIGURE 4. (A) Preoperative anterior segment view of a 6-year-old girl with total bullous exudative retinal detachment, which can easily be seen behind the lens. TDSRF together with PPV, endolaser, and cryotherapy to telangiectatic areas combined with anti-VEGF injection at the end of the surgery resulted in complete reattachment of the retina maintained through the 18 months follow-up. (B) At the last visit, retina was attached completely, together with widespread subretinal fibrosis and subfoveal nodule. Blue arrows indicate the subretinal fibrosis area. PPV = pars plana vitrectomy; TDSRF = transscleral drainage of subretinal fluid.

secondary to ablative treatments. Daruich and associates[19] reported that TRD could contribute to recurrence of ERD also and lead to poor visual outcomes. It is possible that vitreous acts as a scaffold for the formation of ERMs and that its removal may prevent TRD formation in the long term. Vitrectomy also removes all VEGF and inflammatory cytokines from the vitreous, which may explain the long-term effect. However, lensectomy, internal drainage, and formation of

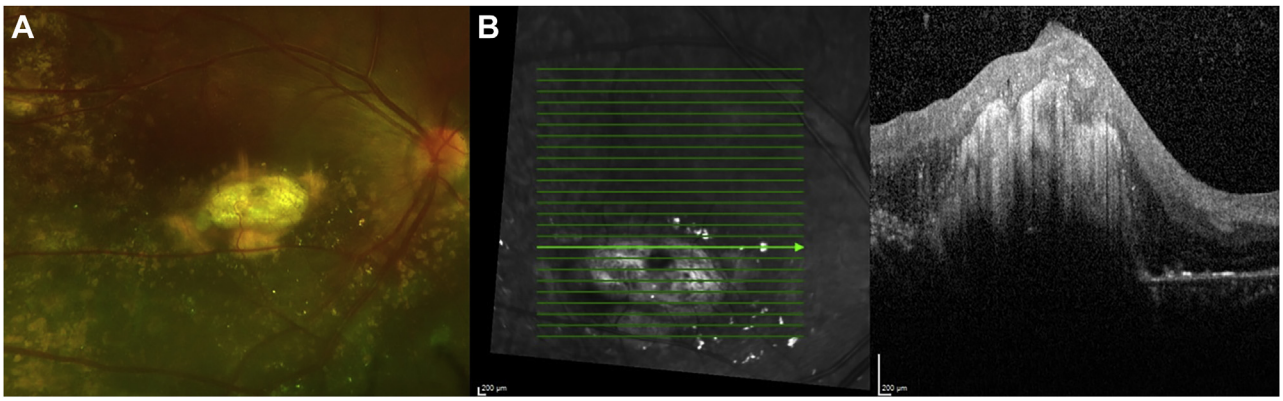

FIGURE 5. (A) Subfoveal nodule, fundus imaging. (B) Optical coherence tomography imaging.

retinal breaks should definitely be avoided during vitrectomy because these may lead to loss of the advantages of PPV in advanced Coats disease.

PPV without drainage was performed by Suesskind and associates[20] in 13 patients with advanced Coats disease, and they reported that 77% of patients achieved retinal reattachment after the surgery. Although there are a few case series reporting favorable outcomes of PPV with internal drainage and tamponade in advanced Coats disease,[19,20] internal drainage retinotomy is usually considered a cause of proliferative vitreoretinopathy and rhegmatogenous retinal detachment. A transscleral method to drain the subretinal exudates and fluid seems to be a safer method that maintains the retinal integrity and decreases the need for long-term vitreous tamponade.

Previous studies evaluating the factors affecting the outcomes of Coats disease revealed that younger presentation (<3 years) was associated with more advanced disease, poor visual outcomes, and a greater need for enucleation.[11,21,22] Similarly, our study showed that younger patients tended to have more advanced disease and poor visual outcomes along with a higher risk of amblyopia. However, the need for further surgical and ablative treatments was similar in patients younger and older than 3 years in this study.

Although relatively successful outcomes have been reported with PPV or TDSRF alone as an adjunct to ablative therapies, knowledge about the efficacy of combined PPV and TDSRF is limited to a single case in the medical literature.[23] The present study is the first to evaluate the effectiveness of combined PPV and TDSRF by comparing it with TDSRF alone as an adjunct to ablative therapies. This study showed a higher rate of surgical success, as well as a lower need for anti-VEGF and laser therapies in the combination group. TDSRF is important for ablative procedures to be more effective in highly elevated ERD. The addition of prophylactic PPV to the surgical procedure could play a protective role against subsequent development of TRD after ablative therapies. The lesser need for further laser and anti-VEGF therapies in TDSRF combined with PPV could be explained by the removal of both VEGF molecules and the vitreous scaffold in vitrectomy, thus decreasing the formation of ERM and tractions that may cause subretinal exudation.

TABLE 3. Complications of Patients with Advanced Coats Disease

	TDSRF	TDSRF + PPV	P^a
Subretinal fibrosis	16 (100)	15 (100)	NA
Subfoveal nodule	15 (93.7)	12 (80)	.275
Tractional RD	4 (25)	–	.058
Seclusio pupillae	–	1 (6.6)	.484
Epiretinal membrane	6 (37.5)	–	**.011**
Hypotonia	–	1 (6.6)	.484

NA = not applicable; PPV = pars plana vitrectomy; RD = retinal detachment; TDSRF = transscleral drainage of subretinal fluid.

Values are n (%). Bold value indicates statistical significance.

[a]Fisher exact test.

Unfortunately, the visual outcomes remain poor in advanced Coats disease, even in patients with favorable anatomical results after surgery, owing to widespread photoreceptor damage and subfoveal nodule formation.[16] The present authors observed visual improvement in a limited number of patients because all the patients in this study had advanced Coats disease at Stage 3 or higher. However, in patients with advanced Coats disease, the primary goal is to stop disease progression, preserve the globe, and thus achieve normal orbital growth until adulthood. Subretinal fibrosis, a common complication of Coats disease, is considered unremarkable because of the low visual expectation. However, ERMs may progress and cause tractional RD leading to anatomical failure and should be followed cautiously. This study showed that the formation of marked ERMs may be prevented in the long term with the addition of PPV to surgery. Hypotonia and seclusio

pupillae, which occurred in 1 patient with stage 4 Coats disease in our study, were not observed in previous studies. No previous studies evaluating surgical outcomes included patients with stage 4 Coats disease. In our study, 2 out of 3 patients with stage 4 Coats disease had TDSRF alone and one had TDSRF combined with PPV; 2 patients who underwent TDSRF alone had retinal reattachment, whereas the remaining patient who underwent combined surgery ended up with failure. Since there was only one case, we cannot make any conclusion depending on this data.

Although this study has some limitations, such as the limited number of patients and a historically controlled design, this study is the first to demonstrate the additional positive effect of PPV on TDSRF and ablative therapy by decreasing the formation of ERMs and the need for further treatments during the follow-up. Designing a prospective study would require time because of the extremely low incidence of Coats disease; however, further randomized controlled studies may help to evaluate the outcomes of these approaches.

In conclusion, although the combination of TDSRF and PPV is a more invasive procedure than TDSRF alone, it appears to be more effective in maintaining retinal reattachment and decreasing the need for additional treatments in the long term, as an adjunct to ablative treatments for advanced Coats disease.

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