Management and surgical outcomes of pediatric retinal detachment associated with familial exudative vitreoretinopathy – Our experience at a tertiary care ophthalmic center in North India

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Purpose: To report the clinical profile, management, and long-term anatomical and visual acuity (VA) outcomes of pediatric macula-off rhegmatogenous retinal detachment (RRD) secondary to familial exudative vitreoretinopathy (FEVR). Methods: This was a prospective, interventional study of 14 eyes of 13 children aged ≤18 years with macula-off FEVR-RRD. The primary outcomes were anatomical reattachment and VA changes. Results: The mean (±SD) age of the study population was 12.14 (±3.23) years (range 6–18 years) with a male preponderance (M:F – 10:3). Of the 14 eyes, 10 underwent vitrectomy with silicone oil injection, while four underwent scleral buckling surgery. Significant improvement in VA was noted at a mean (±SD) follow-up duration of 3.32 (±1.34) years, with the mean (±SD) LogMAR VA improving from 1.42 (±0.48) (Snellen equivalent 2/60; range from 6/36 to counting finger close to face [CFCF]) to 0.6 (±0.31) (Snellen equivalent 6/24; range 6/9–6/36) (P < 0.00001) at the final visit. Successful anatomical reattachment was achieved in 13/14 eyes (92.85%). Screening of the other eye and family members was performed for FEVR and treated with laser photocoagulation when deemed necessary (7/10 contralateral eye; 12/20 siblings; 0/24 parents). Conclusion: To conclude, RRD may arise in eyes with FEVR at a young age and with a male predilection in Indian population. Timely surgical intervention by scleral buckling procedure or vitrectomy, based on the patient profile, can achieve excellent anatomical and VA outcomes. Careful clinical and angiographic screening of the other eye and family members is vital.

Key words: Familial Exudative Vitreoretinopathy, FEVR, retinal detachment, scleral buckling, vitrectomy

Familial exudative vitreoretinopathy (FEVR) is a heritable vitreoretinal disorder characterized by anomalous retinal angiogenesis.[2] This disorder was first described in 1969 by Criswick and Schepens.[1] FEVR has been shown to be bilateral and asymmetric in literature. Incomplete peripheral retinal vascularization and subsequent retinal ischemia are the hallmark features of FEVR.[3] The resultant hypoxia promotes abnormal neovascularization, which, over due course of time, results in secondary complications such as vitreoretinal traction, retinal and disk drag, retinal and vitreous hemorrhage, retinal lipid exudation, falciform retinal folds, and retinal detachments (RDs). [5]

The majority of patients with FEVR remain asymptomatic, and it is not uncommon to miss the diagnosis or erroneously report it as some form of nonspecific vitreoretinopathy. Avascularity of the peripheral retina and abnormal vessels are the most common indicators of FEVR, which are typically missed by fundus examination and necessitate fluorescein angiographic (FA) evaluation.[5] However, the development of RDs is not uncommon in FEVR and can ensue in 21%–64% of these cases. These RDs can be either tractional (TRD), exudative (ERD), rhegmatogenous (RRD), or a combination (CRD) of these. In an Indian series of 61 patients with FEVR and having a mean age of 23.6 years, Shukla et al.[8] reported the surgical outcomes of 14 eyes with RD (10 RRD, three TRD, and one ERD). Management with scleral buckling (SB), vitrectomy, or both led to an excellent reattachment rate of 85.5% with promising visual outcomes.[9] Despite the possibility of visual-threatening complications occurring at a young age, very few studies have evaluated the surgical outcomes of FEVR-associated RD in a pediatric population. Moreover, despite the recent advances in genetics and imaging, the pathogenesis of FEVR remains unclear and the management approach is contentious.

The present study reports the clinical profile and surgical outcomes of macula-off RRD associated with FEVR in an Indian pediatric population.

Methods

This was a prospective, interventional study of consecutive pediatric patients with FEVR having macula-off RRD presenting to a tertiary eye care center in India from December 2014 to July 2022. This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

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2019. The study was conducted in accordance with the tenets of the Declaration of Helsinki and was approved by the Institutional Review Board. Written informed consent for treatment and data collection was obtained from the parents of each patient.

At baseline, the patients underwent a detailed clinical examination by a trained ophthalmologist (S. K.), including best-corrected visual acuity (BCVA) assessment using the Snellen’s visual acuity chart, intraocular pressure (IOP) measurement by Goldmann applanation tonometer, anterior segment evaluation using slit-lamp biomicroscopy, and fundus examination with both slit-lamp biomicroscopy (+90D lens) and indirect ophthalmoscopy (+20D lens). Additionally, all patients underwent a fundus FA with Zeiss fundus camera system (Carl Zeiss Meditec, Dublin, CA, USA) to confirm the diagnosis of FEVR. Optical coherence tomography (CIRRUS 5000, Carl Zeiss Meditec) was also performed. The diagnostic criteria utilized to confirm FEVR were: 1) lack of peripheral retinal vascular development in ≥2 eye, 2) birth at full term or preterm with a disease course incompatible with retinopathy of prematurity (ROP), and 3) variable degrees of nonperfusion, vitreoretinal traction, subretinal exudation, or retinal neovascularization occurring at any age. The inclusion criteria for the study were: 1) a diagnosis of FEVR as described above, 2) presence of an RRD with macula off, for which the patient underwent surgical intervention, and 3) age ≤18 years. Patients were excluded if they had a prior history of any intraocular surgery or if the diagnosis of FEVR was uncertain. Both the eyes of the patient were classified based upon the FEVR staging system proposed by Kashani et al.,[9] namely, stage 1 – a vascular periphery or anomalous intraretinal vascularization without (A) or with (B) exudate or leakage, stage 2 – avascular retinal periphery with extraretinal vascularization without (A) or with (B) exudation or leakage, stage 3 – extramacular RD without (A) or with (B) exudate or leakage, stage 4 – macula-involving RD without (A) or with (B) exudate or leakage, and stage 5 – total RD, which is either open funnel (A) or closed funnel (B).

Surgery
All surgeries were performed by a single experienced surgeon (V. A.) under general anesthesia. SB was done for patients having peripheral retinal proliferation involving less than two quadrants. Silicone tires of suitable dimensions along with a 360° encircling buckle were used in all these eyes. For patients with ≥2 quadrant involvement of fibrovascular proliferation or with its presence at the posterior pole, a primary vitrectomy was undertaken. A standard three-port 23-gauge pars plana vitrectomy was performed. Triamcinolone acetonide was utilized for induction of posterior vitreous detachment. Extensive vitrectomy was undertaken, especially at the vitreous base and around the fibrovascular proliferative tissues. Careful resection and delamination of the fibrovascular tissue were performed to the maximum extent possible. Pneumatic retinal reattachment was achieved followed by endophotocoagulation and/or trans-scleral cryotherapy and silicone oil tamponade. The decision to undertake lensectomy was done in patients having fibrovascular proliferation around the ora and/or at the posterior lens surface.

Follow-up
After the surgery, the patients were evaluated on day 1, week 1, months 1 and 3, and thereafter every 3–6 months depending upon the course of the disease by a trained ophthalmologist (S. K.). Additional surgical interventions were performed as deemed necessary. Silicone oil removal was done 6–9 months following the primary surgery.

The parents and siblings were screened for FEVR and appropriate treatment was offered depending upon the staging.

Demographic and clinical data including age, referral diagnosis, BCVA, FEVR staging, surgical details, any complications, and postoperative data were collected and analyzed.

Statistical analysis
The Statistical analysis was performed using SPSS 23.0 version (SPSS Inc., Chicago, IL, USA). For purposes of statistical analysis, all Snellen visual acuity data were converted to Logarithm of the Minimum Angle of Resolution (LogMAR) values. Continuous variables were described as mean, and variation of each observation from the mean value (standard deviation [SD]) was represented as mean ± 5A or median and interquartile range if they failed to follow a normal distribution. The differences during follow-up were compared with paired t-tests. A P value of < 0.05 was considered to be statistically significant.

Results
Baseline characteristics
We included 14 eyes of 13 patients in this study. Of these, 10 (76.92%) were males and 3 (23.08%) were females. The mean (±SD) age of the children at the time of diagnosis was 12.14 (±3.23) years (range 6–18 years). Bilateral surgery was performed in one patient (7.69%). The mean (±SD) preoperative LogMAR visual acuity in the surgical eye was 1.42 (±0.48) (Snellen equivalent 2/60; ranging from 6/36 to counting finger close to face [CFCF]), while it was 0.3 (±0.2) in the nonsurgical eye (Snellen equivalent 6/12; ranging from 6/9 to 6/36). Table 1 provides the baseline demographic and clinical characteristics, surgical details, and visual and anatomical outcomes of the study population. Table 2 provides the demographic data, preoperative and postoperative details, and the clinical features of family members of the study population.

Clinical Profile
RD was the most common referral diagnosis (8/13; 61.54%), followed by Coats disease (3/13; 23.08%) and ROP (2/13; 15.38%). On fundus evaluation, stage 4A FEVR was noted in six eyes (42.86%), stage 4B FEVR in two eyes (14.28%), and stage 5A in six eyes (42.86%). Simultaneously, the other eye was phthisical in one patient and normal in one patient, while stage 1A FEVR was seen in four eyes, stage 1B in one eye, and stage 2A in five eyes. Laser photocoagulation was done in all eyes having stage 2A and stage 1B FEVR and in one-fourth of eyes with stage 1A FEVR. Among the 20 siblings screened, 12 had stage 2A FEVR, for which they underwent laser photocoagulation. Three out of the 24 parents screened had stage 1 FEVR, which is being observed and closely monitored.

Surgical Management
Out of the 14 eyes with RRD secondary to FEVR, Four eyes underwent SB Silicone oil injection [video 1] (28.57%) and the remaining 10 eyes (71.43%) underwent primary vitrectomy with silicone oil injection [Video 1]. Of these 10 eyes, two eyes had cataractous changes, for which additional cataract extraction with posterior chamber intraocular lens (PCIOL) implantation was also undertaken during the primary surgery. Four eyes that underwent vitrectomy also received preoperative
Table 1: Demographic and clinical characteristics, surgical details, and outcomes of the study population

| Characteristic                          | Number of patients (total 13) | Number of eyes (total 14) |
|----------------------------------------|-------------------------------|--------------------------|
| Age (years)                            |                               |                          |
| Mean (+SD)                             | 12.14 (±3.23)                 |                          |
| Gender                                 |                               |                          |
| Males                                  | 10 (76.92%)                   | 12 (92.85%)              |
| Females                                | 3 (23.08%)                    | 3 (23.08%)               |
| Referral diagnosis                     |                               |                          |
| FEVR                                   | 8 (61.54%)                    | 9 (69.23%)               |
| Coats disease                          | 3 (23.08%)                    | 3 (23.08%)               |
| ROP                                    | 2 (15.38%)                    | 2 (15.38%)               |
| FEVR stage of study eye                |                               |                          |
| Stage 4A                               | 6 (42.86%)                    | 7 (53.85%)               |
| Stage 4B                               | 2 (14.28%)                    | 2 (14.28%)               |
| Stage 5A                               | 6 (42.86%)                    | 7 (53.85%)               |
| Diagnosis/FEVR stage of contralateral eye |                               |                          |
| Stage 1A                               | 4 (33.33%)                    | 5 (38.46%)               |
| Stage 1B                               | 1 (8.33%)                     | 1 (7.69%)                |
| Stage 2A                               | 5 (41.67%)                    | 6 (46.15%)               |
| Normal                                 | 1 (8.33%)                     | 2 (15.38%)               |
| Phthisis                               | 1 (8.33%)                     | 1 (8.33%)                |
| Mean LogMAR BCVA                       |                               |                          |
| Baseline                               | 1.42 (±0.48) (~2/60)          |                          |
| Final                                  | 0.6 (±0.31) (~6/24)           |                          |
| Change in BCVA                         | ⟨0.00001                      |                          |
| Surgical intervention                  |                               |                          |
| Vitrectomy with silicone oil injection | 10 (71.43%)                   | 12 (92.3%)               |
| Additional cataract surgery + PCIOL    | 2 (14.29%)                    | 2 (15.38%)               |
| Scleral buckling                        | 4 (28.57%)                    | 5 (38.46%)               |
| Follow-up duration (years)             |                               |                          |
| Mean (+SD)                             | 3.32 (±1.34)                  | 4.2 (±1.9)               |
| Anatomical retinal reattachment        | 13 (92.85%)                   | 14 (100%)                |

BCVA=best-corrected visual acuity, FEVR=familial exudative vitreoretinopathy, PCIOL=posterior chamber intraocular lens, ROP=retinopathy of prematurity, SD=standard deviation

intraocular anti-vascular endothelial growth factor (anti-VEGF) therapy, since there was presence of active neovascularization on preoperative Fundus Fluorescein Angiography (FFA). All 10 eyes underwent silicone oil removal after 6–9 months.

Visual and Anatomical Outcomes

The patients were followed up for a mean (+SD) duration of 3.32 (±1.34) years, ranging from 1.5 to 5 years. At the final follow-up visit, the mean (+SD) postoperative LogMAR visual acuity improved significantly to 0.6 (±0.31) (Snellen equivalent 6/24; ranging from 6/9 to 6/36) (P < 0.00001). Successful anatomical reattachment of the retina was achieved in all the 13/14 eyes (92.85%) at the final visit. One patient developed sub-silicone oil proliferation leading to retinal re-detachment. No notable surgical/disease-related complications were observed at any of the follow-up visits in the other 13 eyes. Figs. 1–3 provide illustrative case examples of the study population and family members.

Discussion

The current study represents the largest Indian series of macula-off RRD secondary to FEVR in a pediatric population. We demonstrated a significant visual improvement over the long term (mean 3.32 [±1.34] years) in all the eyes with complete anatomical re-attachment, except in one eye (92.85% of cases). Demographically, these patients showed a male preponderance along with variable FEVR involvement in the fellow eye and among parents and siblings.

FEVR is a rare form of vitreoretinopathy that is inherited in nature. Despite this, the inheritance pattern of FEVR is highly variable. In our study, a positive family history from parents was present in 3/13 (23.08%) of patients. This is marginally lower than 37% reported by Ranchod et al. It is pertinent to note that a positive family history encourages and corroborates a diagnosis of FEVR and its absence may not be of much significance.

Clinically, the spectrum of FEVR ranges from completely asymptomatic to several visual-threatening complications such as vitreous hemorrhage and RDs. Since it is inherited in nature, the primary pathogenesis of abnormal retinal vasculature and peripheral retinal ischemia may start very early in life. Hence, it is not uncommon to encounter secondary complications at an early phase of life in these patients. In fact, FEVR is an important cause of juvenile-onset RRD secondary to congenital ocular anomalies. In a study by Pendergast et al., the mean age of the study eyes was only 3.2 ± 4.6 years. In comparison, we had a relatively elder pediatric population with a mean age of 12.14 (±3.23) years. It has been observed that FEVR-associated TRD and ERD usually occur at a younger age, while the development of RRD would take time. This can be due to the severe form of FEVR present in eyes with TRD and ERD, which predisposes them to develop these complications earlier. In contrast, for the development of RRD, these eyes take longer due to gradual vitreous liquefaction and globe enlargement, which is also indicative of a less-severe degree of FEVR. Multiple studies have shown the presence of FEVR-RD among the older population, with the age reported being 16.42 ± 5.48 years by Chen et al., 18.4 ± 9.63 years by Ikeda et al., and 21.8 ± 10.9 years by Yuan et al.

According to the literature, the occurrence of RRD is more common in eastern countries, including Japan and Taiwan. Ikeda et al. reported the surgical outcomes of 28 Japanese eyes with RD in FEVR cases, of which 25 eyes had RRD, whereas only three eyes had TRD. In contrast, multiple studies from the European and American regions have shown a higher preponderance of TRD and ERD in these FEVR patients. In the Indian population, Shukla et al. also noted RRD to be most commonly present in FEVR eyes (20/35 eyes), followed by ERD (10/35 eyes) and TRD (5/35 eyes). Out of these 35 RDs, 14 underwent surgical intervention, including 10 eyes with RRD, and three eyes and one eye with TRD and ERD, respectively. In our series of 14 eyes, all demonstrated the presence of RRD. The exact reason for the varied presentation among the different geographic regions is unknown, but disparities in defining the disease have been proposed to be a possible explanation.

It has been observed that the development of TRD and ERD is not associated with any sex preponderance. However, the presence of RRD is more influenced by the male gender.

Similarly, we too noted the higher rate of RRD in males, comprising three-fourths of the cases. Higher incidence of FEVR in males and/or the presence of some unknown risk factor in males can explain this gender disparity. However,
further genetic studies are warranted to explore this FEVR-RRD relationship with gender in detail.

Our data found significant improvement in visual acuity after the surgical intervention. Concurrently, we had a good anatomical success rate too, with 92.85% of eyes having reattachment of the retina. This is in line with Chen et al.,[5] who too demonstrated excellent retinal reattachment of 95.8% in their series of 24 eyes, and Huang et al.,[13] who achieved anatomical success in 91.67% of eyes undergoing encircling SB with cryotherapy for FEVR-RRD. In other studies, involving a combination of FEVR-RDs, including TRD, ERD, and RRD, marginally lower reattachment rates have been achieved, including 83.9% by Yamane et al.[9] and 83% by Shukla et al.[8] Sen et al.[14] performed a retrospective analysis of 44 eyes of 38 patients with FEVR-RRD undergoing surgical correction. They too reported a slightly lower reattachment rate of 85.7% in 14 eyes undergoing SB and 83.8% in eyes undergoing vitrectomy. This can be due to the advanced nature of the disease in these eyes with the presence of extensive ischemia and secondary proliferative tissues. The primary objective of surgical management in FEVR is to relieve the anteroposterior and tangential tractions and address the avascular retina tissue.[16] The intraocular VEGF load can be reduced by performing a thorough vitrectomy and peripheral retinal tissue ablation by laser photocoagulation.[10] Inducing a posterior vitreous detachment and completing the vitrectomy are critical to relieve the traction onto the retina.[16] Additional usage of SB is vital to mitigate the additional peripheral traction.[10] Depending upon the type and location of the RD and the extent of fibrovascular proliferation, the operating surgeon can decide regarding the kind of surgical approach, SB or vitrectomy. Huang et al.[13] evaluated the surgical outcomes of eyes with only stage 3A or 4A FEVR that were managed by encircling SB with cryotherapy. In contrast, since the current study included eyes having stage 4A, 4B, and 5A FEVR, the surgical

Figure 1: Representative image of an 8/M with FEVR-macula-off RRD with peripheral retinal avascularity and neovascularization seen on the fundus photos (a–c), which was also confirmed on fluorescein angiography (d–f). The patient underwent successful scleral buckling with complete anatomical reattachment (g) and peripheral scarring and disease regression (h). FEVR = familial exudative vitreoretinopathy, RRD = rhegmatogenous retinal detachment.
| Patient | 1 | 1 | 2 | 3 | 4 | 5 | 6 |
|---------|---|---|---|---|---|---|---|
| Eye     | 1 | 2 | 3 | 4 | 5 | 6 | 7 |
| Age (years) | 12 | 12 | 18 | 18 | 15 | 14 | 11 |
| Sex | M | M | M | M | M | M | M |
| Referral diagnosis | Coats disease | Coats disease | RD | ROP | RD | Coats disease | RD |
| Preoperative BCVA | FC3M | FC4M | 6/36 | 6/60 | CFCF | FC2M | CFCF |
| FEVR stage | 5A | 5A | 4A | 4A | 4A | 4A | 5A |
| Treatment | PPV + SOI; SOR | PPV + SOI; SOR | PPV + SOI + cataract extraction PCIOL; SOR | SB | PPV + SOI; SOR | SB | PPV + SOI; SOR |
| Postoperative final BCVA | 6/24 | 6/24 | 6/9P | 6/12 | 6/36 | 6/18 | 6/18 |
| Contra lateral eye status | 5A | 5A | Phthisis | FEVR 2A | FEVR 1A | FEVR 2A | FEVR 2A |
| Contra lateral eye BCVA | FC3M | FC4M | NA | 6/9 | 6/9 | 6/12 | 6/9 |
| Contra lateral eye treatment | PPV+SOI; SOR | PPV+SOI; SOR | NA | Peripheral laser | Observation | Peripheral laser | Peripheral laser |
| Siblings | 2 - FEVR; Laser | 2 - FEVR; Laser | 2-1 had FEVR; Laser | 2 - Normal | 3-2 had FEVR; Laser | Normal | 4-3 had FEVR; Laser |
| Parents | Normal | Normal | Normal | Normal | NA | Normal | Normal |
| Follow-up (years) | 5 | 5 | 2 | 3 | 1.5 | 1.5 | 25 |
| Patient | 7 | 8 | 9 | 10 | 11 | 12 | 13 |
| Eye | 8 | 9 | 10 | 11 | 12 | 13 | 14 |
| Age (years) | 6 | 10 | 17 | 10 | 12 | 13 | 12 |
| Sex | M | M | M | F | F | F | M |
| Referral diagnosis | ROP | RD | Coats disease | RD | RD | RD | RD |
| Preoperative BCVA | 6/36 | CFCF | CF2M | CFCF | 6/60 | CF1M | 5/60 |
| FEVR stage | 4A | 5A | 4B | 5A | 4A | 4B | 5A |
| Treatment | SB | PPV + SOI; SOR | PPV + SOI; SOR | PPV + SOI + cataract extraction PCIOL; SOR | SB | PPV + SOI; SOR | PPV + SOI; SOR |
| Postoperative final BCVA | 6/9 | CF5M | 6/36 | FC3M | 6/18 | 6/24 | 6/24 |
| Contra lateral eye status | FEVR 1A | FEVR 2A | Normal | FEVR 1B | FEVR 1A | FEVR 2A | FEVR 1A |
| Contra lateral eye BCVA | 6/9 | 6/12 | 6/9 | 6/24 | 6/9 | 6/36 | 6/12 |
| Contra lateral eye treatment | Observation | Peripheral laser | NA | Peripheral laser | Observation | Peripheral laser | Peripheral laser |
| Siblings | None | 1 - FEVR; Laser | 1 - FEVR; Laser done | Normal | 1 - FEVR; Laser | None |
| Parents | Normal | Father - stage 1A | Normal | Normal | Normal | Father - stage 1A |
| Follow-up (years) | 4 | 4 | 3 | 2 | 2.5 | 3 | 5 |

**Notes:**
- BCVA=best-corrected visual acuity, CFCF=counting finger close to face, F=female, FC=finger counting, FEVR=familial exudative vitreoretinopathy, M=male, PCIOL=posterior chamber intraocular lens, PPV=pars plana vitrectomy, RD=retinal detachment, ROP=retinopathy of prematurity, SB=scleral buckling, SOI=silicone oil injection, SOR=silicone oil removal
management included both SB and vitrectomy depending upon the clinical profile. Additionally, while Huang et al.\textsuperscript{[13]} included patients with a varied spectrum of age, we evaluated only the pediatric population.

The role of intravitreal anti-VEGF therapy as an adjuvant for treating the vascular and exudative characteristics of FEVR is controversial. Based on the pathogenesis, the anti-VEGF molecules can be helpful in these eyes. However, certain aspects of this treatment, namely, the unforeseen ocular and systemic adverse events and the probability of worsening the fibrovascular traction, make this option less alluring. Against this background, we offered intravitreal anti-VEGF therapy to only four eyes (24.57\%) of our series, which illustrated active neovascularization on preoperative FFA. Nonetheless, future prospective studies are needed to better understand the pathogenesis of FEVR and the role of anti-VEGF therapy in its management.

The limitations of the present study are the limited sample size, inclusion of patients with only FEVR-RRD, and lack of genetic analysis for the patients and their parents and siblings. Absence of eyes with FEVR-TRD and FEVR-ERD in the study cohort may account for the better anatomical outcomes, since these forms of RDs can potentially be trickier to manage. Also, although our mean follow-up period was considerably good (3.32 years), the long-term prognosis can alter in view of known progression of disease after initial stability. Despite these limitations, the current series represents the largest cohort of pediatric macula-off FEVR-RRD in Indian eyes. Although Shukla et al.\textsuperscript{[8]} and Sen et al.\textsuperscript{[14]} have reported larger Indian series on the surgical management of FEVR, their study cohort differed from that of the current study. While the current study included eyes with only macula-off RRD secondary to FEVR, Shukla et al.\textsuperscript{[8]} and Sen et al.\textsuperscript{[14]} treated eyes with both RRD and TRD. Moreover, both these studies included patients from all age groups, ranging from 2 to 55 years (mean 23.6 years) by Shukla et al.\textsuperscript{[8]} and from 5 months to 51 years (median 12 years) by Sen et al.\textsuperscript{[14]} In contrast, the current study evaluated only pediatric eyes (<18 years). Further long-term prospective studies with genetic analysis are needed to better understand their role in the pathogenesis of FEVR and associated RD.

**Conclusion**

In conclusion, FEVR can give rise to RRD at an early age with male gender predisposition in an Indian population. A strong suspicion of FEVR in pediatric eyes with a meticulous clinical and angiographic evaluation of both eyes can bring us to the correct diagnosis and appropriate treatment. Our study shows that timely surgical intervention, either by SB procedure or vitrectomy, is highly effective in achieving anatomical and functional success over the long term.

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**Conflicts of interest**

There are no conflicts of interest.

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