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# Characteristics of Retinal Breaks and Surgical Outcomes in Rhegmatogenous Retinal Detachment in Familial Exudative Vitreoretinopathy

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***Purpose:*** To determine the characteristics of retinal breaks and surgical outcomes in eyes with a rhegmatogenous retinal detachment (RRD) with familial exudative vitreoretinopathy (FEVR).

***Design:*** Retrospective, noncomparative case series.

***Participants:*** Thirty-seven patients (46 eyes) with a RRD in FEVR.

***Methods:*** The medical records were reviewed and the types, directions, and positions of the retinal breaks and surgical outcomes were analyzed.

***Main Outcome Measures:*** Fundus examinations, including ophthalmoscopy, fluorescein angiography, and RetCam imaging (Natus Medical Incorporated, Pleasanton, CA).

***Results:*** The retinal breaks were identified as tears in 12 eyes, atrophic holes in 24 eyes, tears and atrophic holes in 2 eyes, dialysis-related in 1 eye, a retinal break in the ora serrata in 1 eye, and unidentified in 6 eyes. Most retinal breaks (86.1%) were identified only in the temporal retina. Most tears (85.7%) were observed on the demarcation line, whereas atrophic holes were identified both on the demarcation line (53.8%) and the avascular retina (42.3%). The representative tears were almond-shaped, which differs from the typical horseshoe-shaped tears. Scleral buckling was performed as the initial surgery in 37 eyes and resulted in reattachment in 35 eyes (94.6%). Vitrectomy with or without scleral buckle was performed for eyes with more complex RRD in FEVR and resulted in reattachment in 5 of 9 eyes (55.6%). In total, reattachment was achieved in 40 of 46 eyes (87.0%). There was a history of stage 1A or 2A FEVR in 45 eyes and a history of stage 2B FEVR in 1 eye.

***Conclusions:*** Our data clarified the types, directions, and positions of the retinal breaks and the effectiveness of scleral buckling as the first surgical choice for treating RRDs in FEVR. *Ophthalmology Retina 2017;■:1–6 © 2017 by the American Academy of Ophthalmology*

*Supplementary material available at www.ophthalmologyretina.org.*

Familial exudative vitreoretinopathy (FEVR) is a hereditary disorder characterized by insufficient retinal vascular growth similar to retinopathy of prematurity (ROP).[1,2] Clinical complications vary and include retinal dragging, radial retinal folds, vitreous or retinal hemorrhage, exudative changes, and retinal detachment (RD),[3,4] the last of which is a serious complication leading to visual disruption that occurs in 10% to 30% of eyes with FEVR.[5–7]

The surgical outcomes of RD, including rhegmatogenous RDs (RRDs) and tractional RDs, have been studied by multiple researchers,[2,7–9] and we previously reported the surgical outcomes in tractional RDs in FEVR.[10] In contrast, few studies have focused on RRDs in FEVR. Existing data suggested that scleral buckling is effective as the first surgical choice, and vitrectomy should be performed independently or in combination in refractory cases.[8,11] The purpose of this study was to clarify the characteristics of retinal breaks and surgical outcomes in a large series of RRDs in FEVR.

## Methods

The institutional review board of the National Center for Child Health and Development approved the study protocol, which adhered to the tenets of the Declaration of Helsinki. The legal guardians of all patients provided informed consent.

We retrospectively reviewed the medical records (January 2002 to January 2017) in our hospital of patients who were clinically diagnosed with FEVR and in whom a diagnosis of RRD was suspected. We excluded patients whose medical records had missing data or who had an RRD with radial retinal folds because that scenario indicates original and severe retinal deformity accompanied by complex vitreoretinal traction and various locations of the retinal breaks related to the folds. In addition, simple classification of the retinal folds was difficult in the current study. The collected data included the patient age, gender, best-corrected visual acuity (BCVA), fundus examinations, surgical records, follow-up period, and family history of FEVR. We used the BCVA data from just before surgery and after at least 1 year postoperatively. The decimal BCVA was converted to the logarithm of



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the minimum angle of resolution (logMAR) BCVA. Hand motions, light perception, and no light perception visual acuities (VAs) were converted to the values of 2.7, 2.8, and 2.9, respectively, as reported previously.[12] Fundus examinations including fluorescein angiography were conducted using funduscopy, the TRC-50EX fundus camera (Topcon Corporation, Tokyo, Japan), and the RetCam imaging system (Natus Medical Incorporated, Pleasanton, CA). From the fundus examinations and surgical records, we obtained the data from the preoperative laser photocoagulation (LPC) treatment; the degrees of RD; the presence of retinal dragging and proliferative vitreoretinopathy (PVR); the number of surgeries; the type, position, and direction of the retinal breaks; the surgical methods; and the surgical outcomes. The diagnosis of FEVR was established based on the clinical features of the vascular abnormalities with an avascular area in the peripheral retina and no history of prematurity or oxygen supplementation. No genetic testing was performed for diagnosing FEVR. Although the diagnosis of a RRD should be established based on the identification of causative retinal breaks, we could not identify the causative retinal breaks in some patients in whom RRD was strongly suspected from the clinical features (e.g., retinal pigment epithelium cells in the vitreous, clinical course, the form of RD, and exclusion of exudative or tractional RD). These patients were considered to have a RRD and were included in the current study.

The main outcome measure of this study was the retinal reattachment rate. We determined that the optimal time at which to judge the surgical outcome of retinal reattachment was at least 3 months after the last surgery. The secondary outcome was the result of analysis of the types, directions, and positions of the retinal breaks. For the initial surgery, encircling with cryopexy was the first procedure. Encircling was performed using a No. 506 silicone sponge or a No. 240 silicone sponge and a No. 277 silicone tire that was knotted using a No. 270 silicone sleeve. Cryopexy was performed to treat the causative retinal breaks and the degenerated retina. In addition, the subretinal fluid was drained in patients with a bullous RD, and air or sulfur hexafluoride (SF6) gas was injected when a retinal break was insufficiently sealed. When the sealing of retinal breaks by encircling was considered too difficult because of retinal breaks in the posterior pole; substantial degenerated vitreous adhering tightly to the retinal breaks; the presence of multiple retinal breaks that were extensive or in an anteroposterior direction, and/or the presence of PVR; other retinal surgeries, such as radial buckling, vitrectomy, lensectomy, LPC, and air or SF6 gas tamponade, were performed in addition to encircling or independently. When vitrectomy was performed, the core vitreous was removed first, and great care was taken to remove the peripheral vitreous, which adhered tightly to the vascular and avascular areas. The vitreous around the retinal breaks also was removed carefully. Lensectomy was performed if needed to remove vitreous in the peripheral retina more completely.[10] Fluid−gas exchange and air or SF6 gas tamponade were performed in all cases in which vitrectomy was performed. In patients who had had 2 or more surgeries, vitrectomy with additional methods was the first choice. In severe cases, perfluorocarbon and/or silicone oil tamponade was considered. One surgeon (NA) performed all surgeries.

## Results

Forty-six eyes of 37 patients (30 males, 7 females) were studied, including 9 patients who were affected bilaterally. There was a history of stage 1A or 2A in 45 eyes and a history of stage 2B in 1 eye (case 5) before the RRD developed according to the criteria of FEVR.[4] The patient age at the time of diagnosis of RRDs ranged from 3.8 to 20.9 years (mean ± standard deviation [SD], 10.1±3.6). The follow-up periods from the initial surgery ranged from 0.3 to 21.3 years (mean ± SD, 5.8±4.8). A family history positive for FEVR was found in 10 patients (27.0%). Fluorescein angiography had been performed in 17 eyes of 14 patients before or during development of a RRD.

Retinal reattachment was achieved ultimately without silicone oil tamponade in 40 (87.0%) of 46 eyes. Five cases were treated with silicone oil tamponade, including 2 cases with a retinal attachment, 2 cases with a retinal detachment, and 1 unevaluated case with a vitreous hemorrhage. One remaining case had a total retinal detachment. LPC had been performed preoperatively in 7 eyes due to treatment of stage 2B FEVR in 1 eye (case 5), and treatments for avascular fragile retina without exudate or leakage were performed in 6 other eyes. Cryopexy had not been performed preoperatively in our cases. PVR was accompanied by a RRD in 4 eyes. Before surgery the RDs were total in 11 eyes, partial with macula-off in 23 eyes, and partial with macula-on in 12 eyes. Retinal dragging occurred in 15 eyes, did not occur in 30 eyes, and could not be identified in 1 eye because the patient presented to our hospital with a total RD and could not achieve retinal reattachment postoperatively. Surgery was performed once in 33 eyes, twice in 9 eyes, three times in 3 eyes, and four times in 1 eye (mean ± SD, 1.4±0.7 times). Encircling and cryopexy were the first surgical procedures in 36 eyes, including 5 eyes in which the subretinal fluid was drained, 8 eyes injected with air, 1 eye injected with SF6, and 1 eye with radial buckling. Radial buckling without encircling, cryopexy, and LPC was performed in 1 eye. Vitrectomy with LPC and air or SF6 tamponade was performed in 3 eyes, including 1 eye in which lensectomy was performed. Vitrectomy and encircling with LPC, air or SF6 tamponade, and lensectomy were performed in 3 eyes. Vitrectomy, encircling, and radial buckling with LPC and air or SF6 tamponade were performed in 3 eyes, including 2 eyes that had undergone lensectomy. Including eyes that underwent 1, 2, and more surgeries, 10 severely affected or refractory eyes underwent vitrectomy with lensectomy accompanied by additional therapies; among these, 6 eyes did not achieve reattachment. These conditions and surgical outcomes are summarized in Table 1. We measured the logMAR BCVA preoperatively in 42 eyes and after at least 1 year from the last surgery in 41 eyes; the logMAR BCVA changed from 1.0±0.7 to 0.8±1.0.

Retinal breaks were categorized as tears in 12 eyes, atrophic holes in 24 eyes, tears and atrophic holes in 2 eyes, dialysis-related in 1 eye, a retinal break in the ora serrata in 1 eye, and unidentified in 6 eyes. We summarized the types, directions, and positions of the tears and atrophic holes in 36 eyes, which were all components identified in the current study (Fig 1). Most retinal breaks occurred only in the temporal retina (31/36 eyes, 86.1%). Most tears were in the temporal area (13/14 eyes, 92.9%) and on the demarcation line between the avascular and vascular areas (12/14 eyes, 85.7%). The fundus examinations and intraoperative findings confirmed that tears occurred circumferentially due to tight adherence of the vitreous anterior and posterior to the tears, which resulted in a characteristic almond shape (Fig 2). Most atrophic holes also were in the temporal area (22/26 eyes, 84.6%) but both on the demarcation line (14/26 eyes, 53.8%) and the avascular area (11/26 eyes, 42.3%). Figure 2 shows representative fundus photographs of tears and atrophic holes. The detailed clinical data from each case are shown in supplemental Table S1 (available at www.ophthalmologyretina.org).