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FAMILIAL EXUDATIVE VITREORETINOPATHY: PREDISPOSING FACTORS FOR PROGRESSION

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Familial exudative vitreoretinopathy (FEVR) is characterized by incomplete peripheral retinal vascularization due to defects in retinal angiogenesis, leading to peripheral ischemia and subsequent complications such as neovascularization and tractional retinal detachment. While milder forms may remain asymptomatic and exhibit findings such as peripheral retinal avascularity, increased vascular tortuosity, or venous–venous anastomoses, advanced stages are characterized by more severe manifestations, including retinal folds and retinal detachment (1-5). Prophylactic laser photocoagulation (LFC) is commonly employed in early stages (1–2), and vitrectomy (VRC) is often necessary in advanced stages (3–5); unlike retinopathy of prematurity (ROP), which follows a more predictable course, FEVR exhibits a progressive nature that can evolve over months or years, regardless of initial treatment (2,6). Although the concept of progression in FEVR has been recognized since the disease was first described, a clear, standardized definition is lacking. In most studies, progression is defined as worsening of disease stage or increased extent of retinal detachment (1,2,6,7). Due to its rarity, comprehensive studies identifying predictive risk factors for disease progression remain scarce. The aim of this study is to longitudinally evaluate treated FEVR patients to identify clinical or demographic features that may predict long-term progression.

We retrospectively reviewed the charts of all consecutive patients diagnosed with FEVR who had at least 3 years of follow-up between 2004 and 2022. Progression was defined as: (1) worsening of the disease stage, (2) increase in vitreoretinal traction, or (3) increase in exudation. Comparisons of baseline features between groups with and without progression were performed to determine features associated with higher risk.

A total of 191 eyes from 120 patients met the inclusion criteria. Of the 191 eyes, 40 (21%) showed progression after treatment, with a mean follow-up of 78.8 months (range: 36–240 months). Of the exam findings studied, the presence of telangiectatic vascular network and stage B were found to be associated with the development of progression in multivariable analysis.

FEVR patients with telangiectatic vascular network and exudates are more prone to progression and should be followed closely and treated more aggressively.

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