Criswick and Schepens coined the name familial exudative vitreoretinopathy (FEVR) in 1969. The condition appears to have various ways of expressing itself. Usually the eye demonstrates temporal dragging of the retina, producing a loss of acuity over several weeks. Many cases have subretinal exudates, and a fluorescein angiogram can document the extensive peripheral nonperfusion present in almost 90% of the eyes. Other retinal signs are detachments, peripheral cystoid degeneration, neovascularization, and vitreous bands. FEVR is inherited in an autosomal dominant fashion with a high degree of penetrance. There are many mild forms, and an individual may have a mild form in one eye and a severe form in the fellow eye. Retinopathy of prematurity (ROP) should be ruled out before FEVR is considered. Often the severe form of FEVR resembles Stage V ROP. More than 80% of patients have myopia. Color vision and electrophysiological testing tends to be normal. The entire family should be examined when a child presents with a picture of FEVR. Treatment is aimed towards the specific problems, such as photocoagulation or cryopexy of neovascular lesions. Histopathology demonstrates neovascularization (retinal and iris) fibrovascular membranes and exudates.

Selected Reading

FIGURE 13-1.
(A) Pseudoxotropia in the right eye. Note the extreme temporal dragging of the retina. The macula was in a falciform fold, and acuity was at the level of hand motions. (B) The fellow eye did not have the temporal dragging but did have peripheral membranes with peripheral cystoid changes.

FIGURE 13-2.
Fibrovascular proliferative changes in the periphery of a patient with FEVR. The peripheral exudation can be readily seen.