Figure 24-1  Schematic of the retina of the right eye (RE) and left eye (LE), showing zone borders and clock-hour sectors used to describe the location of vascularization and extent of retinopathy. Solid circles represent borders of zones I–III, and dashed circles represent borders of posterior zone II (2 disc diameters beyond zone I). The LE illustration shows a hypothetical example of examination findings, representing approximately 3 clock-hours of stage 1 disease in zone II (the single line documents the presence of stage 1 disease). (Courtesy of Chiang MF, Quinn GE, Fielder AR, et al. International Classification of Retinopathy of Prematurity, 3rd ed. Ophthalmology. 2021;128(10):e51–e68. With permission from Elsevier.)

Figure 24-2  Staging of retinopathy of prematurity (ROP). A, Stage 1 ROP. The demarcation line has no height. B, Stage 2 ROP. The demarcation line has height and width, creating a ridge. C, Stage 3 ROP. Ridge with extrafoveal fibrovascular proliferation. D, Stage 4A ROP. Partial detachment of the retina not involving the fovea. E, Stage 4B ROP. Partial detachment of the retina involving the fovea. F, Stage 5 ROP. Total retinal detachment. (Part A courtesy of Daniel Weaver, MD; part B courtesy of Andrea Molinari, MD; part C reproduced with permission from Lueder GT. Pediatric Practice Ophthalmology. McGraw-Hill Medical; 2011:232. Permission conveyed through Copyright Clearance Center, Inc.; parts D–F courtesy of R.V. Paul Chan, MD, and Michael F. Chiang, MD.)
**CLINICAL PEARL**

Clinically, the temporal edge of zone I is visible with a 25.00 D or 28.00 D lens, with the other edge of the field of view on the nasal optic nerve head margin.

*Plus disease* (Fig 24-3) and *pre–plus disease* (Fig 24-4) are a continuum of marked arteriolar tortuosity and venous dilation of retinal vessels as assessed in zone I (see Table 24-1).

The term *aggressive posterior ROP (AP-ROP)* was previously used to describe a severe, rapidly progressive form of ROP in posterior zone I and posterior zone II. However, because this aggressive form of ROP can occur outside the posterior zones and in larger preterm infants, particularly in countries with limited resources, the ICROP3’s new preferred term is *aggressive ROP (A-ROP)*, which reflects the variable location of disease. A-ROP is associated with plus disease out of proportion to the stage of ROP present. In addition, A-ROP does not progress in the typical fashion (ie, through stages 1, 2, then 3), and stage 3 often

**Figure 24-3** Wide-angle fundus photos demonstrating the spectrum of plus disease as defined by the International Classification of Retinopathy of Prematurity, 3rd edition (ICROP3). The vascular abnormalities are assessed in the central retina within the region of zone I. **A**, Plus disease with notable venous dilation and arterial tortuosity. The plus disease is out of proportion to visible peripheral findings, suggesting flat neovascularization (stage 3, arrows). **B**, Severe plus disease, with dilation and tortuosity of both arteries and veins. **C**, Severe plus disease. Note the presence of ill-defined posterior flat stage 3 (arrows), which, combined with severe plus disease, is typical of aggressive ROP (A-ROP). (Courtesy of Chiang MF, Quinn GE, Fielder AR, et al. International Classification of Retinopathy of Prematurity, 3rd ed. Ophthalmology. 2021;128(10):e51–e68. With permission from Elsevier.)