

# VEIN OCCLUSIONS: CLINICAL CLUES FOR IDENTIFICATION



Help patients avoid permanent vision loss by knowing how to identify these vascular disorders.

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**R**etinal vascular occlusions are the second most common retinal vascular disorder, following only diabetic retinopathy.<sup>1</sup> Risk factors, signs, and symptoms of retinal vein occlusion (RVO) vary based on the type of vein occlusion and the age of the patient. Although identification of RVO is fairly simple in the acute presentation, eye care practitioners must often become retina detectives to properly diagnose and manage a chronic, quiescent RVO. Whether an RVO is active or quiescent, accurate identification is necessary to ensure proper management of any underlying systemic etiology.

This article differentiates the types of RVO to aid practitioners in identifying even the most subtle retinal changes in order to better collaboratively manage their patients with RVOs.

## GET TO KNOW YOUR RVOs

There are three types of RVO: branch RVO (BRVO), central RVO (CRVO), and hemiretinal vein occlusion (HRVO). Each type has different characteristics in its acute and inactive phases. The long-term visual



Figure 1. Large symptomatic superotemporal BRVO with significant macular edema.

prognosis of RVO depends on macular involvement and ischemia, which can lead to neovascularization.

## BRVO

BRVO describes an occlusion of one of the major retinal venules, most frequently secondary to arteriolar compression at an arteriovenous crossing. In the acute presentation of a BRVO, only one quadrant of the retina will exhibit venous dilation or tortuosity, intraretinal hemorrhages, and cotton wool spots (Figure 1). The most frequent location of a BRVO is the superotemporal quadrant (nearly 50%), which has a higher number



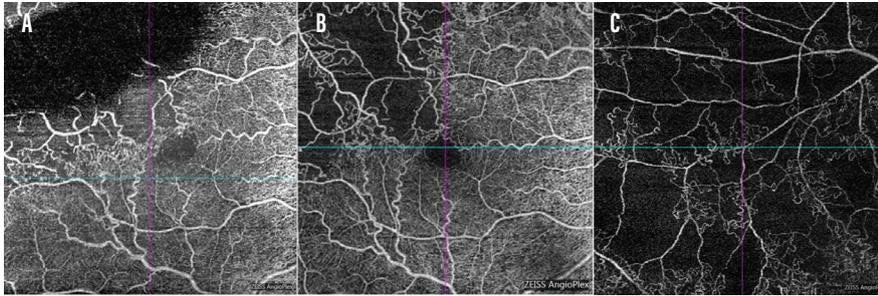


Figure 2. Superficial OCTA taken 3 weeks after the patient seen in Figure 1 was diagnosed with ischemic BRVO (A). Collateral vessels and > 5 DD of capillary dropout can be seen, although partially obscured by superotemporal intraretinal edema. Same patient at 17 months after diagnosis of ischemic BRVO (B); superior arcade at 17 months (C).

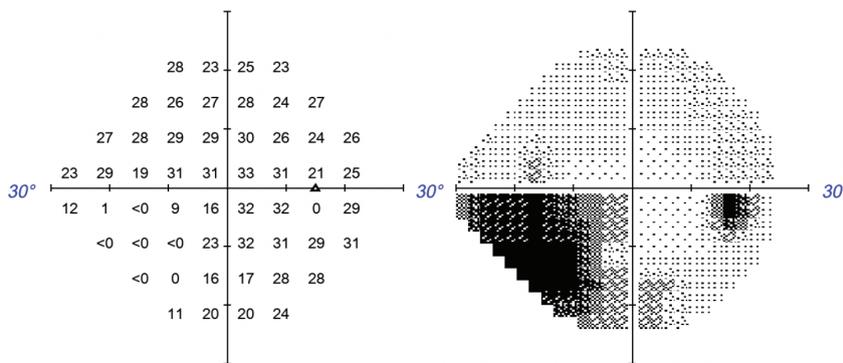


Figure 3. Visual field shows inferior nasal arcuate defect corresponding to superotemporal BRVO shown in Figure 2.

of arteriovenous crossings than the other quadrants.<sup>2</sup> Other locations for BRVO include inferotemporal and nasal. Cystoid macular edema (CME)

may be present, depending on the size and location of the BRVO.

*Retinal capillary dropout* is an important vascular change to look

for in determining whether a BRVO is ischemic in nature. Eyes with an area of retinal nonperfusion greater than 5 disc diameters (DD) are at increased risk of retinal neovascularization, which occurs at the border of perfused and nonperfused retina. In a BRVO, the most common types of neovascularization noted are retinal and optic disc. Capillary dropout can be difficult to assess with fluorescein angiography (FA) when dense intraretinal hemorrhages are present. Therefore, FA is typically not performed until about 3 months after the occlusion is noted. OCT angiography (OCTA) is another option for evaluating capillary dropout that can be fairly accurate when employed early enough (Figure 2). BRVOs classified as nonischemic rarely convert to ischemic status.

In an eye with a quiescent BRVO, both small and large collateral vessels will be evident (Figure 2). The larger retinal venules (from occluded vein to nonoccluded vein) will cross the horizontal raphe. Smaller intraretinal collateral vessels may be noted in areas of capillary nonperfusion that represent remodeling of the retinal capillaries.<sup>3</sup> OCT will demonstrate thinning of the retinal nerve fiber layer (RNFL) after about 6 months.<sup>4</sup> This thinning can easily be confused with glaucomatous RNFL thinning if the subtle collateralization is overlooked. RNFL thinning and associated visual field loss (Figure 3) can also mimic nonarteritic anterior ischemic optic neuropathy (NAION). A BRVO can be differentiated from NAION by evaluating the optic nerve for pallor and evaluating peripapillary capillary perfusion using OCTA.

### CRVO

CRVO describes an occlusion of the central retinal vein as it traverses the lamina cribrosa or slightly thereafter. In the acute presentation of a CRVO, all four quadrants of the retina exhibit venous dilation and

## AT A GLANCE

- ▶ There are three types of retinal vein occlusion (RVO): branch, central, and hemiretinal.
- ▶ The most common causes of vision loss in patients with BRVO are cystoid macular edema, macular ischemia, and vitreous hemorrhage secondary to retinal neovascularization.
- ▶ In the acute presentation of CRVO, all four quadrants of the retina exhibit venous dilation and tortuosity, intraretinal hemorrhages, and cotton wool spots.
- ▶ In an HRVO with central RVO, the superior and inferior portions of the vein coalesce posterior to the lamina cribrosa.



## RVO FACTS

**FACT** ⇒ ALL THREE TYPES OF RVO HAVE SIMILAR RISK FACTORS.

# 65

More than half of patients with RVO are older than 65 years and have some type of systemic vascular disorder.

# 48

In nearly 48% of patients with RVO, hypertension is identified as a systemic association.

**FACT** ⇒ IMPORTANT RISK FACTORS FOR RVO INCLUDE HYPERLIPIDEMIA, ARTERIOSCLEROSIS, DIABETES, AND SMOKING.<sup>1</sup>

1. O'Mahoney PR, Wong DT, Ray JG. Retinal vein occlusion and traditional risk factors for atherosclerosis. *Arch Ophthalmol*. 2008;126(5):692-699.

tortuosity, intraretinal hemorrhages, and cotton wool spots. Cystoid macular edema is likely, and optic nerve head engorgement may be observed.

Two clinical tests can provide an excellent sense of visual prognosis in acute CRVO: VA and pupillary testing. Patients with ischemic CRVO will exhibit a VA of less than 20/200 and a relative afferent pupillary defect. Ischemic CRVOs have a poor visual prognosis.

As in BRVO, retinal capillary dropout is an important vascular change for determining retinal perfusion status. Eyes with greater than 10 DD capillary dropout are at increased risk of retinal neovascularization. Potential locations for neovascularization in CRVO include the retina and optic disc, but commonly neovascularization is found in the anterior segment (iris and angle), where it can lead to severe neovascular glaucoma. Capillary dropout can be difficult to assess with FA when dense intraretinal hemorrhages are present; therefore, at this time it is best to classify the CRVO as of indeterminate stage, unless VA or pupillary tests indicate nonperfusion.

Unlike BRVOs, which often have stable perfusion status, about one-third of CRVOs will convert to ischemic status within a few months. FA or OCTA can be used to help determine the amount of retinal ischemia.

In an eye with a quiescent CRVO, large collateral vessels, also known as optociliary shunt vessels (from occluded vein to choroidal vein) may be evident at the optic nerve head. These shunt vessels are not unique to CRVOs, however; they can also be seen in severe glaucoma, optic nerve sheath meningioma, or compressive optic neuropathies. In compressive lesions, optic nerve pallor will also be apparent.

### HRVO

Like CRVO, HRVO can occur when there is an occlusion of the central retinal vein. In an HRVO with CRVO, however, the superior and inferior portions of the vein coalesce posterior to the lamina cribrosa. Therefore, when an occlusion occurs, only the inferior or superior half of the retina will display venous distention,

intraretinal hemorrhages, and cotton wool spots. CME is likely in both inferior and superior HRVO but is more visually significant in superior HRVO.

Because HRVO is uncommon, the amount of capillary dropout that determines whether it is ischemic or nonischemic is not well established. Thus, in regard to treatment protocols, HRVO is generally considered a BRVO. Neovascularization that occurs in ischemic HRVO can be posterior (optic nerve or retina) or anterior (iris or angle). Quiescent HRVO will consist of both CRVO and BRVO, and both small and large retinal collateral vessels and optociliary disc shunts can be seen.

### SUSSING OUT SYMPTOMS

Patients with acute RVOs may or may not be symptomatic, and symptoms will depend on the location of the RVO and whether or not the macula is involved. Patients with a superotemporal BRVO are the most likely to report symptoms. The most common causes of vision loss in patients with BRVO are CME

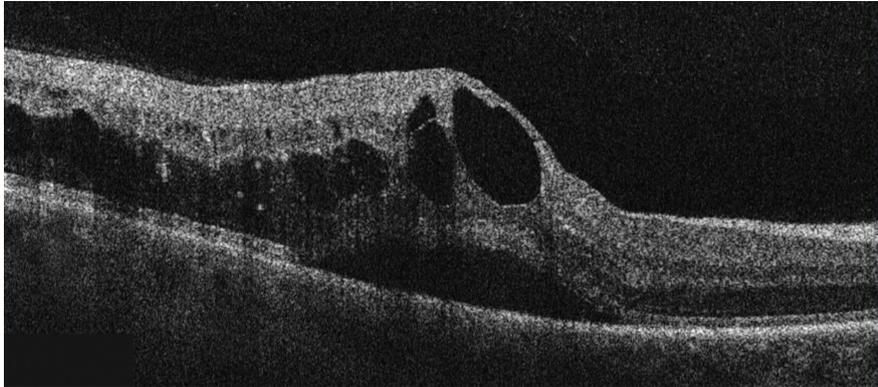


Figure 4. In this OCT image, BRVO with CME and subfoveal detachment are seen.

(Figure 4), macular ischemia, and vitreous hemorrhage secondary to retinal neovascularization.

Patients with a CRVO will present with sudden painless loss of vision secondary to CME, ischemic maculopathy, vitreous hemorrhage secondary to posterior segment neovascularization, and/or neovascular glaucoma. Patients with acute CRVO should be monitored monthly with gonioscopy to rule out anterior segment neovascularization, as this can lead to a difficult-to-manage, painful, blind eye.

With all RVOs, it's important to provide prompt treatment to patients who have reduced vision secondary to CME, as chronic CME can lead to pigmentary degeneration and outer retinal atrophy resulting in long-term vision loss.

### WHO IS AT RISK?

Understanding who is at risk of RVO will help to ensure that any systemic conditions that contribute to the RVO are attended to (see *RVO Facts*). For patients who do not fit the typical presentation of an RVO, it is important to determine the underlying etiology.

Elevated plasma homocysteine is a well-known risk factor for thrombotic events, including RVO.<sup>5</sup> Low levels of vitamin B6 and folic acid have also been identified as risk factors for RVO.

Other associated systemic risk factors for BRVO include arterial hypertension and peripheral vascular disease. Ocular associations of BRVO include arteriovenous nicking and focal arteriolar narrowing as seen in hypertensive retinopathy.

Systemic risk factors for CRVO that have been discussed but remain controversial include increased blood viscosity caused by protein C or S deficiency or factor V Leiden mutation.<sup>1</sup> Glaucoma is an ocular association of CRVO, and inflammatory and infectious etiologies are also known to contribute to CRVO and should therefore be ruled out.

### BE SURE TO GET THE COMPLETE PICTURE

Atypical patients with any type of RVO require further examination to determine whether a systemic condition has contributed to the RVO. Recommended evaluations include

blood pressure measurement and bloodwork. In some cases an MRI may also be indicated. Bloodwork may include complete blood count with differential, lipid panel, prothrombin time, partial thromboplastin time, clotting factors (C and S), factor V Leiden, homocysteine, erythrocyte sedimentation rate, C-reactive protein, antinuclear antibodies, rheumatoid factor, syphilis (FTA-ABS or rapid plasma reagin), and antiphospholipid antibodies. Keep in mind that blood tests can be expensive, especially those that are genetic in nature (eg, factor V Leiden, clotting factors).

Putting all the pieces together (clinical findings, patient symptoms, patient demographics) allows clinicians to narrow a differential diagnosis (eg, glaucoma, NAION, compressive lesions) and make appropriate clinical recommendations for patients with RVO. In some cases, these actions by the primary eye care provider could save a patient's life. ■

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■ Financial disclosure: None