Systemic Hypertension

Effects of Systemic Hypertension on the Eye and Vision

- Hypertensive retinopathy
- Hypertensive choroidopathy
- Hypertensive optic neuropathy
- Central and branch retinal vein occlusion
- Retinal macroaneurysm
- Vitreous hemorrhage
- Subconjunctival hemorrhage
- Secondary effects
  - Carotid disease—retinal arterial emboli
  - Cerebrovascular accident—visual field loss
  - Ischemic optic neuropathy
  - Oculomotor cranial nerve palsies
  - Cataract, glaucoma, age-related macular degeneration

Hypertensive Retinopathy

Hypertensive retinopathy is usually asymptomatic. In severe hypertension, however, painless loss of vision may occur due to the development of vitreous or retinal hemorrhages, retinal edema, retinal vascular occlusion, serous retinal detachment, choroidal ischemia, or optic neuropathy.

Fundus Features

- Focal or diffuse narrowing of retinal arterioles
- Increased vascular tortuosity (Fig. 27.1)
- Arteriovenous nicking (nipping) (Fig. 27.2)
- Retinal hemorrhages (Fig. 27.3)
  - Flame-shaped hemorrhage
  - Dot and blot hemorrhage
  - Preretinal hemorrhage
- Microaneurysms
- Hard exudates, macular star
- Cotton-wool spots (Figs. 27.4 and 27.5)
- Abnormality of vascular reflex
  - Copper-wire reflex (Fig. 27.6)
  - Silver-wire reflex (Fig. 27.7)
- Intraretinal microvascular abnormalities
- Retinal edema (Fig. 27.8)
- Serous retinal detachment—occurs secondary to hypertensive choroidopathy in patients with very severe hypertension or eclampsia (Fig. 27.9)
- Elschnig spots—small to medium-size hyper- and hypopigmented patches representing chorioretinal scarring due to prior choroidal infarction (Figs. 27.10 and 27.11)
III  The Retina in Systemic Disease

- Siegrist streaks—linear configurations of hyperpigmentation that have a pathogenesis similar to that of Elschnig spots.
- Optic disc edema (Fig. 27.12)
- Optic atrophy
- Vitreous hemorrhage

![Fig. 27.1](image1) Increased retinal vascular tortuosity in a patient with chronic systemic hypertension.

![Fig. 27.2](image2) Arteriovenous nicking (arrows) is narrowing of the retinal vein at the site of crossing of an artery. It results from thickening of the retinal arterial wall in chronic hypertension.
Fig. 27.3  Grade 3 hypertensive retinopathy—multiple dot and blot hemorrhages, flame-shaped hemorrhages, and microaneurysms in a patient with uncontrolled hypertension.

Fig. 27.4  Grade 3 hypertensive retinopathy—multiple flame-shaped hemorrhages, cotton-wool spots, hard exudates, and area of retinal edema (arrow).
Fig. 27.5  Cotton-wool spots, flame-shaped hemorrhages, and arteriovenous nicking in a patient with hypertension.

Fig. 27.6  Copper-wire reflex, vascular tortuosity, and blurring of optic disc margin in a patient with severe hypertension.
**Fig. 27.7**  Silver-wire reflex, hard exudates, and hemorrhages are present in a patient with a history of malignant hypertension. Scars of retinal laser photocoagulation are present.

**Fig. 27.8**  (A) Retinal edema, multiple cotton-wool spots, and fine hard exudates in a 32-year-old man presenting with blurring of vision and blood pressure of 220/140 mm Hg. (B) Area of retinal edema is delineated with dotted line. (C) Hard exudates developed in a radial pattern (macular star) 6 weeks after lowering of blood pressure and resolution of the retinal edema.
Fig. 27.9  Patchy area of choroidal infarct (arrow) with overlying serous retinal detachment in a patient with severe hypertension.

Fig. 27.10  (A) Yellow, placoid, subretinal lesions indicating choroidal ischemia in a patient with severe systemic hypertension. (B) Pigmented atrophic scars developed in the areas of choroidal ischemia 2 months after the initial presentation.
Fig. 27.11  “Elschnig spots”—small to medium-size hyper- and hypopigmented patches representing scarring due to prior hypertension-induced choroidal infarction.

Fig. 27.12  Grade 4 hypertensive retinopathy (“accelerated” or “malignant” hypertension). Optic disc is hyperemic, and edematous with blurring of the margin. Vascular tortuosity and retinal hemorrhages are present.
Classification of Hypertensive Retinopathy

Several classification systems for hypertensive retinopathy have been described, including that by Keith, Wagener, and Barker in 1939. The current conventional classification is:

- Grade 0—no changes
- Grade 1—arteriovenous nicking (nipping), increased vascular tortuosity, and minimal arteriolar narrowing
- Grade 2—arteriolar narrowing with focal irregularities
- Grade 3—retinal hemorrhages, microaneurysms, hard exudates, cotton-wool spots (plus grade 2)
- Grade 4—optic disc swelling (plus grade 3); also termed malignant or accelerated hypertension (Fig. 27.13)

![Fig. 27.13](A) Grade 4 hypertensive retinopathy in a patient with blood pressure of 240/150 mm Hg. Optic disc edema, multiple hemorrhages, and early “macular star” are present. Peripapillary circumferential folds indicate presence of retinal edema and sub-retinal fluid. (B) Prepapillary gliosis, circumferential folds, vascular tortuosity, and hemorrhages 4 months after blood pressure was brought under control.

Differential Diagnosis

- Diabetic retinopathy
- Central or branch retinal vein occlusion
- Papilledema
- Carotid artery disease
- Radiation retinopathy
- Vasculitis
- HIV retinopathy
- Neuroretinitis
- Purtscher-like retinopathy
- Other forms of retinal vasculopathy
Central and Branch Retinal Artery Occlusion (CRAO and BRAO)

Symptoms
- Acute, painless, monocular loss of vision (rarely bilateral)
  - Partial (quadrantic or hemifield loss)—branch retinal artery occlusion (BRAO)
  - Central or paracentral scotoma—small macular branch artery occlusion
  - Complete—central retinal artery occlusion (CRAO)
  - Tunnel vision—central retinal artery occlusion with sparing of cilioretinal artery
  - Transient—amaurosis fugax

Fundus Features
- Early
  - Opaque, grayish-white retina (Fig. 27.14)
  - Thickened edematous retina
  - Obscured retinal pigment epithelium (RPE) and choroidal features (Fig. 27.15)
  - Cherry-red spot at the macula—due to the normally thin anatomy of the retinal layers in the foveal region that allows the reddish light reflex from the intact choroidal vasculature to stand out in contrast to the grayish-white appearance of the surrounding thicker and more opaque retina (Fig. 27.16)
  - Minor retinal hemorrhages
  - Retinal artery embolus—an embolus is visible in 20 to 30% of cases
  - Narrow arterioles
  - Segmented “box-car” arterioles
- Late
  - Disappearance of retinal edema and cherry-red spot
  - Attenuated retinal arterioles
  - Atrophic, featureless retina
  - Visible retinal artery embolus
  - Subtle, diffuse, or patchy pigment mottling
  - Pale optic disc—optic atrophy
  - Iris neovascularization—develops in 15% of cases with CRAO. The patient is at highest risk during the first 3 months.
Fig. 27.14  Small branch retinal artery occlusion—yellow-white clouding of retina and cotton-wool spots in the distribution of a small retinal artery. The fovea is spared.

Fig. 27.15  Cloudy swelling of the superior retina in a patient with occlusion of the upper retinal arteries. “Box-carring” of some of the arteries indicates sluggish blood flow. Emboli are visible within the vessels at the optic disc and the superotemporal periphery.
Fig. 27.16 Central retinal artery occlusion. The retina is diffusely edematous and cloudy. “Cherry-red” spot appearance at the macula results from transmission of choroidal color through the relatively thin retina in this region. An embolus is visible in a small retinal arteriole inferior to the fovea.

Causes of Retinal Artery Occlusion

- Embolus
  - Platelet emboli (Fig. 27.17)—are soft, gray or yellowish, and conform to the shape of the blood vessel. They usually originate from an ulcerating atheromatous plaque within the carotid artery and can cause amaurosis fugax. They may be seen moving along the retinal vasculature.
  - Cholesterol emboli (Fig. 27.18)—Hollenhorst plaques are flat, yellow crystalline deposits that are commonly found at the bifurcation of the retinal arteries. They are often asymptomatic. They signify atheromatous disease of the carotid artery.
  - Calcific emboli (Fig. 27.19 and 27.20)—Calcific emboli have a pearly white appearance, are larger, and tend to lodge in the larger retinal arteries around the optic disc. Calcific emboli usually occlude the blood flow and are visually symptomatic.
  - Septic emboli—can cause white-centered hemorrhages (Roth spots)
  - Fat emboli—Purtscher-like retinopathy
  - Amniotic fluid emboli—Purtscher-like retinopathy
  - Talc emboli—intravenous drug abuse
- Hypercoagulable state
- Retinal vasculitis
- Sickle cell disease—more frequently affects the smaller peripheral arterioles
- Vasospasm—migraine
- Dissecting aneurysm of internal carotid artery and its smaller branches
- Ocular trauma
- Severe elevation of intraocular pressure
  - Acute glaucoma
  - Intraocular surgery
  - External pressure on the eye (e.g., during general anesthesia)
- Severe elevation of orbital pressure
  - Acute retrobulbar hemorrhage
- Other associations
  - Mitral valve prolapse
Fig. 27.17  Extensive platelet emboli conforming to the shape of the blood vessels.

Fig. 27.18  Cholesterol emboli (Hollenhorst plaques) are flat, yellow crystalline deposits that are commonly found at the bifurcation of the retinal arteries (arrows). The most common source of this type of embolus is an atheromatous lesion of the carotid artery.
Fig. 27.19 A calcific embolus is impacted at the bifurcation of the superonasal artery. Note attenuation of the artery and dark-colored blood (representing stagnation) distal to the embolus. Cotton-wool spots (CWSs) are present.

Fig. 27.20 A calcific embolus and platelet emboli are present in the arteries over the optic disc. This patient also had a history of central retinal vein occlusion as evidenced by the collateral vessels on the disc and venous sheathing.
Sources of Retinal Emboli

- Carotid artery atheromatous plaque
- Cardiac valve abnormalities
- Cardiac defects
- Atrial myxoma
- Bacterial endocarditis/septicemia/fungemia
- Intravenous drugs (paradoxical emboli)

Amaurosis Fugax

Amaurosis fugax is due to a transient embolic phenomenon within the retinal circulation, resulting in a transient, monocular, painless loss of vision that usually lasts less than 1 hour. Amaurosis fugax is often associated with other forms of transient ischemic attacks (TIAs) occurring within the carotid artery distribution. During the attack, ophthalmoscopy may show bright yellow cholesterol emboli. The source of these emboli is often carotid atheromatous plaque.

Differential Diagnosis of Cherry-Red Spot in Macula

- Central retinal artery occlusion
- Sphingolipidoses
  - Gangliosidoses (Tay–Sachs disease and Sandhoff disease)
  - Niemann–Pick disease types A to D
  - Metachromatic leukodystrophy
  - Farber disease
- Mucopolysaccharidoses
  - Hurler disease
- Mucolipidoses; sialidoses

Ophthalmic Artery Occlusion

- Severe visual loss, often to the level of light perception or worse. This degree of visual loss is rare in isolated central retinal artery occlusion with retention of choroidal or optic nerve perfusion.
- Pale opaque retina, no cherry-red spot (because choroidal circulation is also diminished)
- Ocular hypotony
- Serous retinal and choroidal detachment
- Bone-spicule pigmentary changes in late stages
- Iris neovascularization

Central and Branch Retinal Vein Occlusion (CRVO and BRVO)

Symptoms

- Acute onset, painless loss of vision corresponding to the area of distribution of the occluded vessel. Loss of vision may not occur in BRVO.
Fundus Features

- Early (Figs. 27.21 and 27.22)
  - Superficial retinal hemorrhages—severe cases have a “blood and thunder” appearance
  - Microaneurysms
  - Retinal edema
  - Cotton-wool spots
  - Capillary telangiectasia
  - Blurring of optic disc margin or disc edema
  - Serous retinal detachment—severe cases

- Late (Figs. 27.23 and 27.24)
  - Above signs
  - Hard exudates
  - Collateral (shunt) vessels at the optic disc (Fig. 27.25)
  - Venous sheathing
  - Retinal or optic disc neovascularization
  - Vitreous hemorrhage
  - Tractional retinal detachment
  - Iris neovascularization—more common in CRVO than in BRVO or CRAO; most cases occur within the first 3 months

Fig. 27.21 Branch retinal vein occlusion (BRVO)—superficial flame-shaped hemorrhages and deeper intraretinal hemorrhages in the distribution of the occluded superotemporal retinal vein.
Fig. 27.22  Hemorrhagic central retinal vein occlusion (CRVO)—extensive flame-shaped and blot hemorrhages in a “blood and thunder” pattern indicating severe hemorrhagic CRVO. CWSs are suggestive of ischemia, and retinal folds represent edema. The optic disc is swollen and has multiple superficial hemorrhages.

Fig. 27.23  Branch retinal vein occlusion (BRVO), 4 months after the initial presentation in Fig. 27.21. Most of the hemorrhages have resolved. Telangiectasis, residual hemorrhages, venous sheathing, and hard exudates, representing retinal edema and vascular incompetence, are present.
Fig. 27.24  Mild, nonhemorrhagic central retinal vein occlusion (CRVO). Flame-shaped and dot and blot hemorrhages, telangiectasis, and vascular tortuosity are present in all quadrants, suggestive of CRVO. The optic disc is hyperemic and has fine collateral vessels.

Fig. 27.25  Large optic disc collateral vessels are present in this patient with an old CRVO. In contrast to new vessels, collateral vessels do not leak or cause hemorrhage.
Risk Factors for Retinal Vein Occlusion

- Age
- Systemic hypertension
- Cardiovascular disease
- Obesity
- Hypercoagulable states
- Hyperviscosity syndromes
- Diabetes mellitus
- Sickle cell hemoglobinopathies—rare
- Glaucoma
- Retinal vasculitis

◆ Retinal Arterial Macroaneurysm

Retinal arterial macroaneurysms are acquired, abnormal saccular dilatations of retinal arteries. They are often multiple and may be bilateral in 10% of affected individuals. Retinal macroaneurysms often undergo spontaneous sclerosis and closure.

Symptoms

- Acute painless loss of vision due to vitreous or retinal hemorrhage
- Chronic painless loss of vision due to progressive macular edema
- May be asymptomatic

Fundus Features

- Red round vascular lesion, measuring 100 to 500 μm, on or adjacent to a retinal artery (Fig. 27.26). The lesions may be solitary or multiple, and unilateral or bilateral.
- Hemorrhage (Fig. 27.27)
  - Retinal
  - Subretinal
  - Subhyaloid
  - Intravitreal
- Retinal edema
- Hard exudates—often occur in a circinate pattern centered on the macroaneurysm
- Capillary telangiectasis
- Vascular sheathing
- Retinal vascular occlusion

Conditions Associated with Retinal Macroaneurysm

- Systemic hypertension—60%
- Retinal vasculopathies (e.g., Coats disease, Leber military aneurysms)
- Septic emboli
- Idiopathic
Fig. 27.26  Retinal macroaneurysms with surrounding retinal edema and hard exudates. Focal chorioretinal scars of prior retinal laser photocoagulation are present.

Fig. 27.27  Retinal macroaneurysm (MA) causing hemorrhages at various layers, including subretinal (SRH), retinal (RH), and retrohyaloid (RHH) hemorrhages. Presence of fine hard exudates indicates chronic leakage.
Retinal manifestations of carotid artery disease may be due to embolization to the retinal vasculature or to the ocular ischemic syndrome. Retinal arterial embolism is discussed earlier in this chapter. Ocular ischemic syndrome is usually associated with greater than 90% stenosis of the carotid artery.

**Symptoms**
- Gradual onset of visual loss
- Impaired dark adaptation
- Periocular and periorbital pain—pain improves on lying down

**Fundus Features**
- Flame-shaped and blot hemorrhages ([Fig. 27.28](#))—Typically, retinal hemorrhages are prominent in the equatorial region.
- Microaneurysms
- Cotton-wool spots
- Narrowed retinal arteries
- Dilated (but often not tortuous) retinal veins. This contrasts with retinal vein occlusion where the veins are both dilated and tortuous.
- Retinal and optic disc neovascularization

**Other Ophthalmic Features**
- Ocular hypotony
- Anterior chamber flare
- Iris atrophy
- Iris neovascularization

![Fig. 27.28](#) Slow-flow retinopathy due to carotid artery stenosis. The retinal veins are dark and dilated (not tortuous). Multiple intraretinal blot, flame-shaped, and dot hemorrhages are present, along with CWSs.
Hyperlipidemia

Hyperlipidemia is associated with general atheromatous disease and, therefore, with its sequelae in the retina. In addition, severe hyperlipidemia may result in slow retinal blood flow, causing vascular tortuosity, dilatation, and mild retinal hemorrhages. In severe hyperlipidemia, the retinal vessels appear salmon-pink or yellow rather than red (Figs. 27.29 and 27.30).

Fig. 27.29  Salmon-pink and tortuous retinal vessels in a 32-year-old African-American male with severe hypertriglyceridemia.

Fig. 27.30  Xanthelasma of the eyelid skin in a patient with hyperlipidemia.
• Patients with grade IV hypertensive retinopathy are at significantly increased risk of cardiovascular and cerebrovascular morbidity and mortality. They require urgent management of their hypertension.

• In patients with diabetes, poorly controlled hypertension exacerbates diabetic retinopathy. Blood pressure control is an integral part of managing patients with diabetic retinopathy.

• Retinal vein occlusion primarily affects individuals of 50 years of age or older. Young patients with severe or bilateral retinal vein occlusion require further evaluation for possible underlying disease, including hypercoagulable states and systemic vasculitis.