RETINAL DISEASE

Retinal detachment, macular disease, and retinal vascular occlusion are all associated with sudden visual loss. Acute visual loss may develop in any inflammatory process that affects the retina, including infectious chorioretinitis, vasculitides, and idiopathic inflammation. These conditions may be distinguished from other causes of acute visual loss by their ophthalmoscopic findings.

Retinal Detachment

Acute visual loss is a feature of an extensive retinal detachment. Typically, the patient with a retinal detachment (Figure 2.2) complains of flashing lights followed by large numbers of floaters and then a shade over the vision in one eye. A detachment extensive enough to reduce visual acuity usually produces a relative afferent pupillary defect in the involved eye. The diagnosis of an extensive retinal detachment is made by ophthalmoscopy through the dilated pupil. The retina appears elevated, sometimes with folds, and the choroidal background is indistinct. However, the findings may not be obvious, and emergency ophthalmologic consultation is indicated.

Macular Disease

Macular disease reduces visual acuity, but unless the disease is extensive, a relative afferent pupillary defect may not be present. Sudden visual loss or metamorphopsia (a defect of central vision in which the shapes of objects appear distorted) from macular disease is often a sign of bleeding from a neovascular net formed as part of the process of age-related macular degeneration (see Chapter 3). The neovascularization may be identified and treated with laser surgery although progression to significant and permanent visual loss often occurs.

Retinal Vascular Occlusion

Retinal vascular occlusion is a relatively common cause of sudden visual loss and may be transient or permanent. Transient monocular visual loss due to

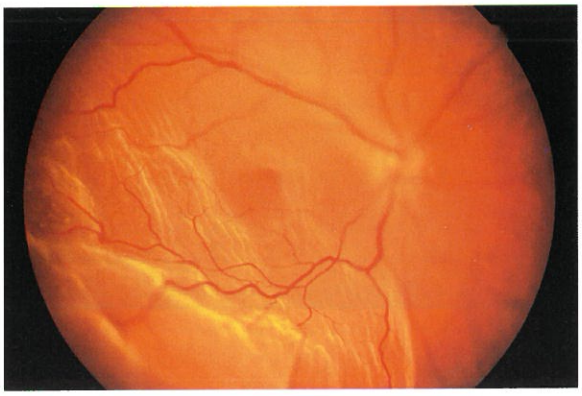


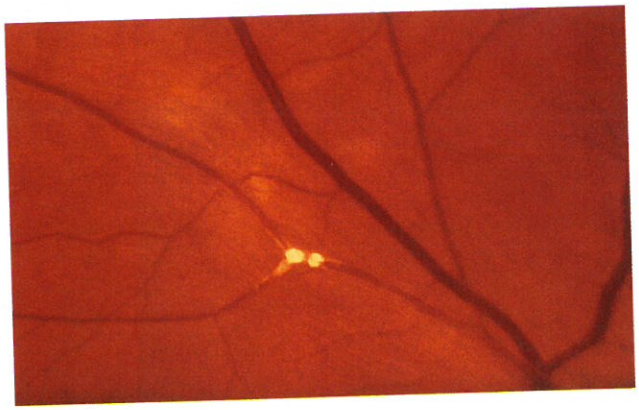
FIGURE 2.2 Retinal detachment. A wide-angle photograph of the fundus reveals folds of retina extending into the macula inferotemporal to the disc. In this photograph, the focus is on the elevated retina, which renders the disc slightly out of focus.

arterial insufficiency is called *amaurosis fugax* and is a very important symptom. In a patient over age 50, the report of visual loss in one eye lasting for several minutes should lead to investigation of the ipsilateral carotid circulation, looking for an atheroma. The valves and chambers of the heart should also be investigated (Figure 2.3), looking for an embolic source causing transient interruption of blood flow to the retina. The evaluation and management of such a patient raises complicated issues, and referral should be made to an ophthalmologist, a neurologist, or a vascular surgeon.

Central Retinal Artery Occlusion (CRAO) Prolonged interruption of retinal arterial blood flow causes permanent damage to the ganglion cells and other tissue elements. CRAO (Figure 2.4) is manifested as a sudden, painless, and often severe visual loss. The ophthalmoscopic appearance depends on how soon after the visual loss the fundus is seen. Within minutes to hours, the only findings may be vascular stasis: narrowing of arterial blood columns and interruption of venous blood columns with the appearance of "boxcarring" as rows of corpuscles are separated by clear intervals.

Some hours after a central retinal artery occlusion, the inner layer of the retina becomes opalescent. The loss of the normal transparency of the retina is most visible ophthalmoscopically where the retina is thickest around the fovea. In the fovea itself, the inner layers are attenuated and the underlying intact choroidal circulation is seen. Pallor of the perifoveal retina stands in contrast to the normal color of the fovea, causing the characteristic "cherry-red spot" of CRAO. (A chronic cherry-red spot is also a feature of storage

FIGURE 2.3 Cholesterol embolus in retinal arteriole. In the elderly, the most common sources of emboli are fibrin and cholesterol from ulcerated plaques in the wall of the carotid artery. The so-called *Hollenhorst plaque* is a cholesterol embolus that lodges at an arterial bifurcation, as shown here. *(Reprinted from* Ocular Manifestations of Systemic Disease. A Slide-Script Program. *San Francisco: American Academy of Ophthalmology; 1996:8.)*



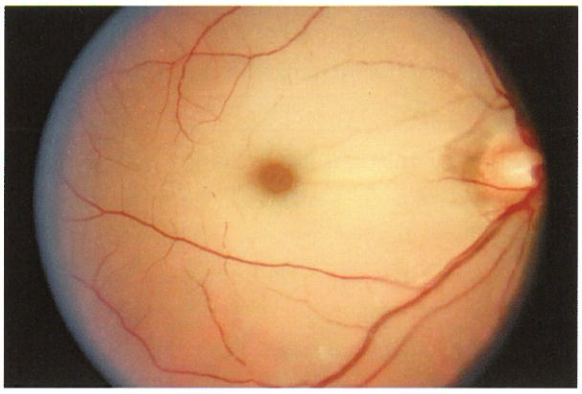


FIGURE 2.4 Central retinal artery occlusion. The retina is opaque, except for the relatively thin area within the macula, producing the "cherry-red spot."

diseases, such as Tay-Sachs disease and some variants of Niemann-Pick disease, in which the ganglion cells become opalescent because of the deposition of intermediate metabolites.)

The optic disc, which is supplied by other branches of the ophthalmic artery, does not swell unless the occlusion is in the ophthalmic or carotid artery, proximal to the origin of the central retinal artery or in the small vessels supplying the disc. The peculiarities of the eye's vascular supply also can explain the possible preservation of some vision in the presence of a complete CRAO. If part of the retina derives its blood supply from the choroidal circulation via a cilioretinal artery, its function is spared. After a CRAO, the retinal edema slowly resolves and the death of the ganglion cells and their axons leads to optic atrophy. Months later, the characteristic ophthalmoscopic appearance is a pale disc in a blind eye.

When ophthalmoscopy reveals an acute CRAO, immediate treatment is warranted unless circulation has already been restored spontaneously. *This is a true ophthalmic emergency*; restoration of blood flow may preserve vision if the occlusion is only a few hours old. Instances are reported in which vision has returned after treatment of an occlusion that has been present for several days. In a blind eye, there is little to lose by aggressive measures, and an ophthalmologist's advice should be obtained emergently.

As an emergency measure, the primary care physician may wish to compress the eye with the heel of the hand, pressing firmly for 10 seconds and then releasing for 10 seconds over a period of approximately 5 minutes. The sudden rise and fall in intraocular pressure could serve to dislodge a small embolus in the central retinal artery and restore circulation before the retinal tissues sustain irreversible damage. An ophthalmologist might employ more vigorous and invasive techniques, such as medications to lower intraocular pressure, vasodilators, and paracentesis of the anterior chamber. Although

most retinal artery occlusions are embolic in nature, central or branch retinal artery occlusion in an elderly patient without a visible embolus should be evaluated for giant cell arteritis.

Branch Retinal Artery Occlusion (BRAO) When only a branch of the central retinal artery is occluded, only a sector of the retina opacifies and vision is only partially lost. The patient will know the moment of vision loss and be able to describe or draw the exact outline of the missing area of vision. A BRAO (Figure 2.5) is more likely to be the result of an embolus than is a CRAO, and a source should be sought. If visual acuity is affected, attempts should be made to dislodge the embolus by ocular massage, as discussed above.

Central Retinal Vein Occlusion (CRVO) The ophthalmoscopic picture of disc swelling, venous engorgement, cotton-wool spots (which appear as small white patches on the retina), and diffuse retinal hemorrhages indicates a CRVO (Figure 2.6). Loss of vision may be severe, although the onset is generally subacute, unlike the dramatic sudden blindness of a central retinal artery occlusion. The fundus picture can be so striking that the description "blood and thunder" is applied. Despite its dramatic appearance, there is no generally accepted acute management, and a CRVO is not a true ophthalmic emergency.

A CRVO is most often encountered in older patients with hypertension and arteriosclerotic vascular disease. Carotid artery occlusion may produce a similar but milder fundus picture. In rare cases, diseases that increase blood viscosity—such as polycythemia vera, sickle-cell disease, and lymphoma-leukemia—induce a CRVO.

The acute hemorrhages and disc swelling resolve with time; however, they may be followed by the development of shunt vessels from the retinal to the choroidal circulation or by ocular neovascularization. The patient with a CRVO needs a general medical evaluation and follow up by an ophthalmologist, who may be able to prevent the late complication of neovascular glaucoma

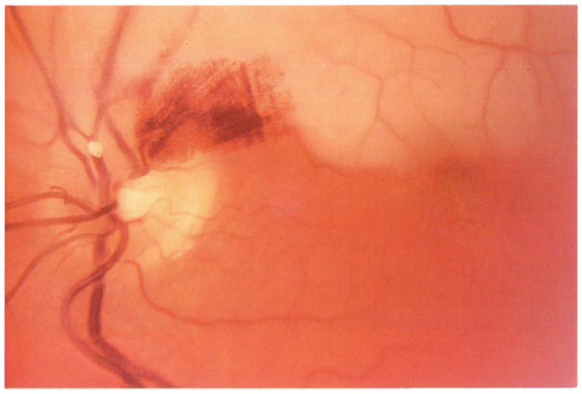


FIGURE 2.5 Branch retinal artery occlusion. Inferotemporal branch retinal artery obstruction. *(Courtesy Cynthia A. Bradford, MD.)*

loss may point to a noninflammatory cause, for example, by a finding of visual field loss in the other eye. Computed tomography or magnetic resonance imaging of the orbits and chiasmal region will identify most compressive lesions, which are potentially treatable with surgery.

Papillitis and Papilledema

Like retrobulbar optic neuritis, papillitis (Figure 2.7) is a subtype of optic neuritis. Specifically, *papillitis* is an inflammation of the optic disc, or papilla. *Papilledema* (Figure 2.8), on the other hand, refers to swelling of the optic disc from increased intracranial pressure; both optic discs are affected. In optic neuritis (either retrobulbar neuritis or papillitis), vision is usually (but not always) significantly decreased, and examination of the pupils will reveal a relative afferent pupillary defect. In papilledema, the visual acuity and the pupillary reflexes are usually normal. In both conditions, fundus examination

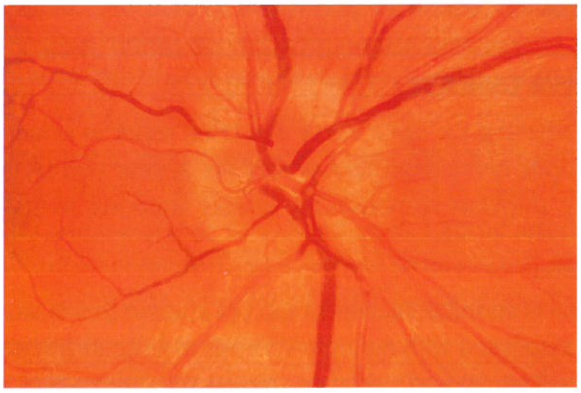


FIGURE 2.7 Papillitis. The disc is swollen, with blurred disc margins. In papillitis, the disc is hyperemic, rather than pale as in ischemic optic neuropathy. Papillitis is usually unilateral. Bilateral papillitis can be differentiated from papilledema based on decreased visual acuity in papillitis.

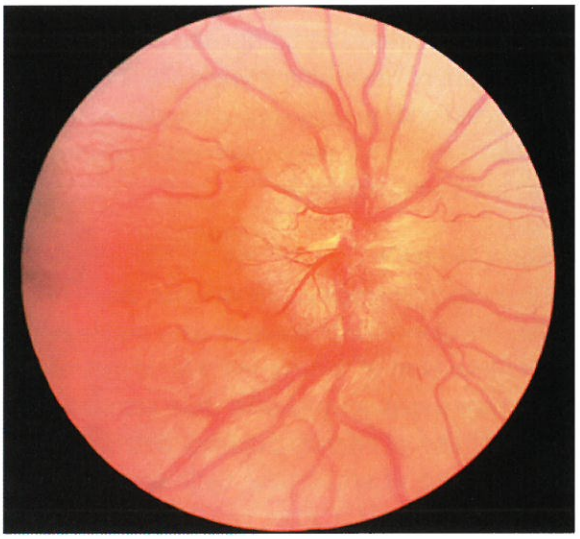


FIGURE 2.8 Papilledema. The optic disc is elevated and the margins are indistinct. There is microvascular congestion on the disc, the retinal veins are dilated, and flame-shaped hemorrhages are present. The appearance in the other eye should be similar.

will reveal blurred optic disc margins, and the optic disc cupping is typically obliterated.

Some patients with acute papilledema complain of momentary blurring or transient obscurations of vision. Although chronic papilledema may lead to loss of vision, most patients with acute papilledema suffer only minor alterations in vision. An emergent brain scan to identify an intracranial mass is indicated. Patients with pseudotumor cerebri will have papilledema without a midline shift on brain scan, and a spinal tap is necessary to document increased intracranial pressure.

Ischemic Optic Neuropathy

Swelling of the disc and visual loss in an older adult are likely to represent a vascular event rather than inflammation. *Ischemic optic neuropathy* (Figure 2.9) is a vascular disorder that presents as a pale, swollen disc, often accompanied by splinter hemorrhages and loss of visual acuity and visual field. The field loss with ischemic neuropathy is often predominantly in the superior or inferior field, a pattern known as *altitudinal.*

Giant Cell Arteritis

The development of acute ischemic optic neuropathy in a patient over age 60 raises the possibility of giant cell, or temporal, arteritis. Patients being considered for the diagnosis of giant cell arteritis (GCA) should undergo a focused review of systems. Common complaints associated with GCA are temporal headache or tenderness, often causing pain while resting on a pillow; scalp tenderness with hair brushing; ear or anterior neck discomfort (carotidynia); fatigue or pain of the tongue or jaw with chewing (jaw claudication); and episodes of transient diplopia or visual loss. Other complaints include anorexia, weight loss, general malaise, and aching/fatigue of the upper arms or legs (polymyalgia rheumatica).

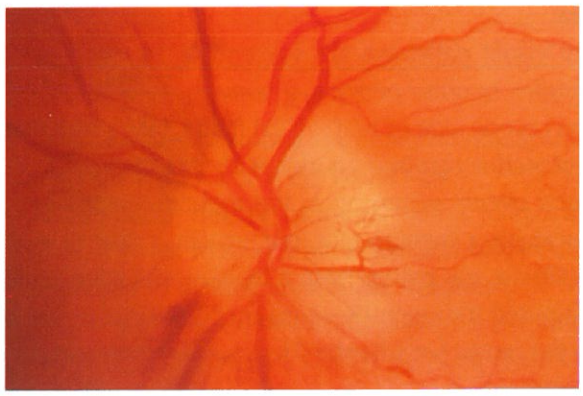


FIGURE 2.9 Ischemic optic neuropathy. This figure shows pale swelling of the optic disc, with associated flame-shaped hemorrhages.

Measurement of intraocular pressure as an independent risk factor is not a valuable means of screening for glaucoma. Glaucoma screenings that include formal (ie, automated, not manual confrontational) visual field analysis and examination of the optic nerve may be beneficial but are costly and require trained personnel. Thus, evaluation of risk factors and examination of the optic nerve by the primary care physician with appropriate referral to the ophthalmologist is the best defense against this silent, potentially blinding disease.

BASIC INFORMATION

This section reviews intraocular pressure (IOP), the types of glaucoma, the optic nerve, and the relationship of IOP and the optic nerve.

INTRAOCULAR PRESSURE

Within the eye is a mechanism for the continuous production and drainage of fluid. This fluid, called *aqueous humor*, is produced by the ciliary body of the eye. Aqueous humor flows through the pupil into the anterior chamber,

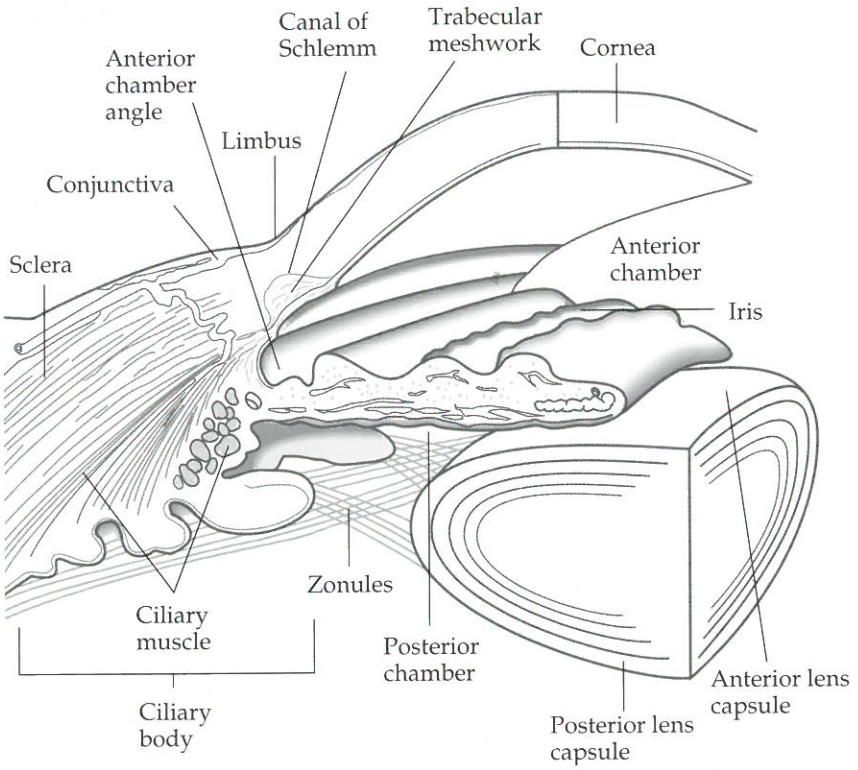


FIGURE 3.3 Cross-section of anterior chamber angle and ciliary body.

where it is drained through the trabecular meshwork to Schlemm's canal (Figure 3.3), and onward to the venous system. Because of some resistance to the flow of aqueous through the trabeculum and Schlemm's canal, pressure is created in the eye. All eyes have an internal pressure.

Intraocular pressure is largely dependent on the ease of flow through the trabeculum and Schlemm's canal. The greater the resistance to flow, the higher the pressure in the eye. Although the eye contains several compartments within it, for purposes of pressure it can be considered a single closed space. Thus, the pressure exerted within the eye is equal over the entire wall of the eye. Most normal eyes have an IOP of 21 mm Hg or less.

TYPES OF GLAUCOMA

In the common, insidious form of glaucoma, the chamber angle remains open. Accordingly, this form of glaucoma is called *open-angle glaucoma.* In rare instances, the trabeculum can become suddenly and completely occluded by iris tissue. This causes an abrupt rise in intraocular pressure known as *acute angle-closure glaucoma* (Figure 4.1) and constitutes an ocular emergency. The abrupt rise in pressure causes symptoms of pain, nausea, and colored halos or rainbows around light. *These symptoms do not occur in open-angle glaucoma.* An acute attack of angle closure usually produces a red, teary eye with a hazy cornea and a fixed, mid-dilated pupil. The eye feels extremely firm to palpation in most cases.

Rather than persistent symptoms, chronic angle-closure glaucoma has intermittent, low-grade symptoms of headache and blurred vision, especially with situations that induce pupillary dilation. Gradual scarring of the drainage angle occurs resulting in elevated IOP.

Several developmental disorders can lead to elevated IOP. Congenital or infantile glaucoma presents as tearing and sensitivity to light secondary to corneal edema from the elevated intraocular pressure. If the pressure remains elevated, the immature eye tissues expand and the eye enlarges (buphthalmos). In contrast, juvenile glaucoma presents later in life and is much more insidious.

Glaucoma may be associated with other ocular disorders, systemic disease, medications, ocular trauma, and inflammation, and may occur following intraocular surgery. In addition, some forms of glaucoma have an identifiable genetic basis, but this accounts for a very small percentage of all patients with glaucoma.

OPTIC NERVE

The optic nerve is composed of more than 1.2 million nerve fibers. These nerve fibers originate in the ganglion cells of the retina, gather in a bundle as

the optic nerve, and carry visual information to the brain. An interruption of these nerve fibers results in damage to vision.

The optic nerve can be seen at its origin by using the ophthalmoscope. At the point of origin, the nerve is called the *optic disc*. The optic disc often has a small depression in it called the *cup of the optic disc*. The size of the cup in normal eyes can vary with the individual. A complete description of the optic disc appears in Chapter 1.

RELATIONSHIP OF IOP AND THE OPTIC NERVE

Intraocular pressure is exerted on all "walls" of the eye, including the optic nerve and its blood vessels. The optic nerve is supplied with blood via branches of the ophthalmic artery, itself a branch of the internal carotid artery. Vascular disease may predispose patients to optic nerve damage. Thus, eye pressure-dependent and eye pressure-independent factors (eg, vascular disease) play a role in the development of glaucoma. Glaucoma is the general term used to describe the progressive optic neuropathy that can lead to blindness if untreated.

This optic neuropathy that is called glaucoma causes visual field loss. Early nerve damage usually causes localized peripheral visual field loss that is often undetectable by the patient, but which can progress to impair central vision and eventually total vision if the disease is not adequately treated. Currently, all treatment regimens are directed toward lowering IOP. Measurement of IOP, evaluation of the optic nerve appearance, and visual field testing play a key role in diagnosis and monitoring of glaucoma.

WHEN TO EXAMINE

Ophthalmoscopy should be part of every comprehensive eye examination. Particular attention should be given to patients who are predisposed to glaucoma, such as elderly individuals or those with a family history of glaucoma. The American Academy of Ophthalmology recommends a glaucoma screening every 2 to 4 years past age 40, as the incidence of the disease increases with age. Because African-Americans have an even greater risk for development of glaucoma, those between ages 20 and 39 should additionally be screened every 3 to 5 years.

HOW TO EXAMINE

Palpation can detect only very hard and very soft eyes; it is totally unreliable in the range of the most common glaucomatous intraocular pressures. Intraocular pressure is best measured via tonometry, which may be performed in any of several ways. Indentation, or Schiøtz, tonometry involves an inexpen-

sive instrument that is simple to use. However, hand-held applanation tonometers are available and give much more reliable readings. Detailed information on various methods of tonometry appears in Chapter 1. The technique of direct ophthalmoscopy, also described in Chapter 1, is particularly useful in assessing the state of the optic disc.

An ophthalmologist evaluating a patient with suspected glaucoma will examine the anterior chamber angle structures using a special contact lens on the topically anesthetized cornea, a technique called *gonioscopy*. In addition, measurement of the central corneal thickness may be important to aid in determining the risk of glaucoma. Thin corneas result in artificially low IOP measurement, and thick corneas measure higher than the actual IOP. The ophthalmologist will also perform perimetry (automated visual field testing). Finally, documentation of the optic nerve appearance for future comparison is performed with stereo disc photography or other forms of optic nerve analysis.

HOW TO INTERPRET THE FINDINGS

The appearance of the optic disc can be described generally in terms of its color and of the size of its physiologic cup (a recognizable central depression within the optic disc). The color of the optic nerve can be important in determining atrophy of the nerve that is due to glaucoma or other causes. Temporal pallor of the optic nerve (Figure 3.4) can occur as a result of diseases that damage the nerve fibers, such as brain tumors or optic nerve inflammation, or in conjunction with glaucomatous cupping.

The term *glaucomatous cupping* refers to an increase in the size of the optic cup relative to the optic disc that occurs in glaucoma. The increase in the cup is due to loss of nerve fibers bundled in the optic nerve. This so-called cup:disc ratio is determined by comparing the diameter of the disc to that of the cup (Figure 3.5). The optic discs generally should appear symmetric between the eyes, and asymmetric cup:disc ratios should arouse suspicion of

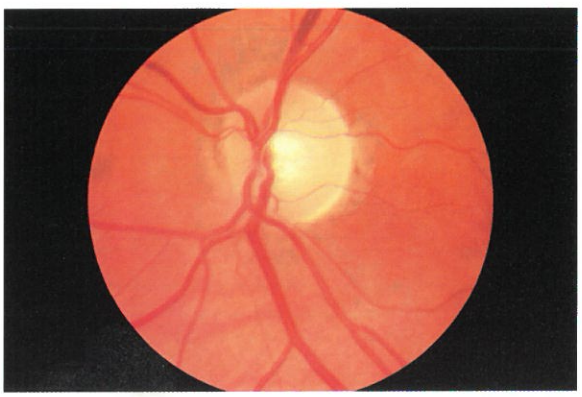


FIGURE 3.4 Temporal pallor of the optic nerve. Diseases that damage optic nerve fibers may result in temporal pallor of the optic nerve. Note the normal nerve color present only on the nasal aspect of the disc.

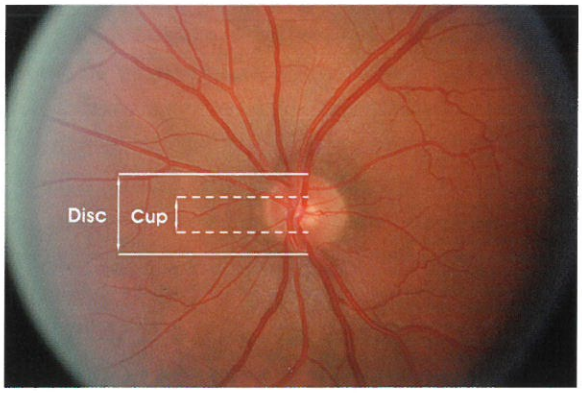


FIGURE 3.5 Cup:disc ratio. In this nondiseased optic disc, the cup is less than one half the diameter of the disc, indicating absent or low level of suspicion of glaucoma.

glaucoma. The larger the cup, the greater the probability of a glaucomatous optic nerve. A cup measuring one half the size of the disc or larger—a cup:disc ratio of 0.5 or more—raises suspicion of glaucoma (Figure 3.6). Disc hemorrhages (Figure 3.7) are also a possible sign of glaucoma. A large cup should be suspected if central pallor of the disc is prominent. Because the cup is a depressed area of the disc, retinal vessels passing over the disc are seen to

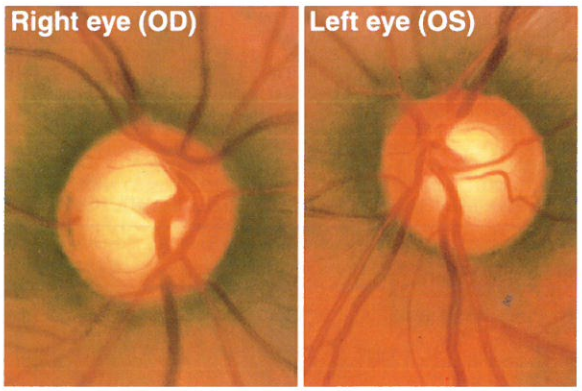


FIGURE 3.6 Glaucomatous cupping. Patient's right eye shows a cup:disc ratio of 0.8 (high level of glaucoma suspicion); the left eye shows a cup:disc ratio of 0.6 (moderate level of glaucoma suspicion). The asymmetry of cup:disc ratios here also raises suspicion of glaucoma.

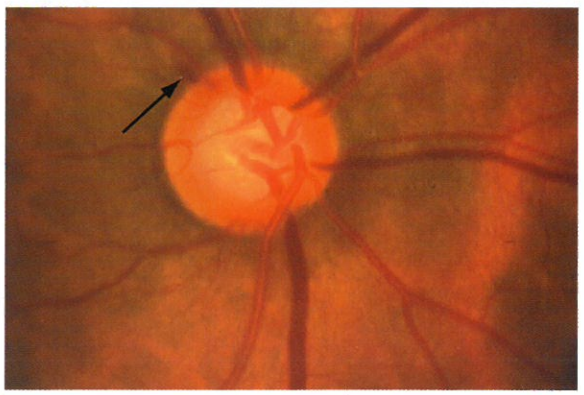


FIGURE 3.7 Disc hemorrhage. A hemorrhage on the optic disc may indicate glaucomatous damage.

bend at the edge of the cup, a useful sign in evaluating cup size. Vessel displacement, then, as well as disc color, should be evaluated in determining the size of the cup (Figure 3.8).

MANAGEMENT OR REFERRAL

Table 3.1 provides a convenient method of analyzing a patient's level of glaucoma risk. A moderate or high level of glaucoma risk warrants referral to an ophthalmologist for further evaluation. In addition, any patient who has one or more of the following conditions should be referred to an ophthalmologist:

- Symptoms of acute glaucoma (Refer immediately; also refer to Figure 4.1.)
- An optic cup diameter one half or more of the disc diameter (ie, a cup:disc ratio of 0.5 or greater)
- Cup:disc asymmetry of more than 0.1 between the two optic nerves (eg, right eye = 0.4, left eye = 0.2)
- Annual ophthalmologic screening for glaucoma in elderly patients and younger African-American patients

For a discussion of systemic side effects of topically administered drugs used in the treatment of glaucoma, see Chapter 9.

CATARACT

RELEVANCE

Cataract may occur as a congenital or genetic anomaly, as a result of various diseases, or with increasing age. Some degree of cataract formation is to be expected in all people over age 70. In fact, age-related cataract occurs in about 50% of people between ages 65 and 74 and in about 70% of those over 75.

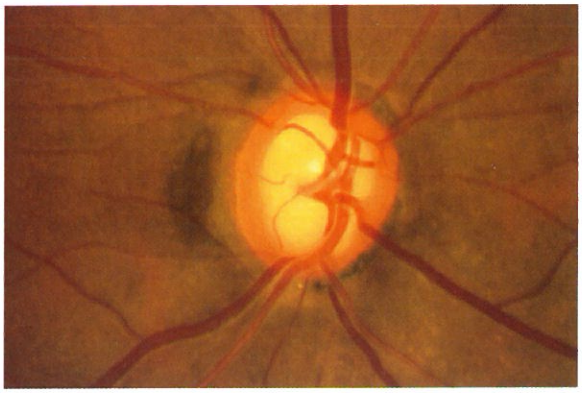


FIGURE 3.8 Glaucomatous optic atrophy. Optic nerve cupping is increased vertically, with a cup:disc ratio of 0.8. Cupping is apparent at the point where the vessels disappear over the edge of the attenuated rim.

TABLE 3.1 Glaucoma Risk Factor Analysis

History-Based Risk Factor Weights

Variable*	Category	Weight
Age	<50 years	0
	50–64 years	1
	65–74 years	2
	75 years	3
Race	Caucasian/other	0
	African-American	2
Family history of glaucoma	Negative or positive in non–first-degree relatives	0
	Positive for parents	1
	Positive for siblings	2
Last complete eye examination	Within past 2 years	0
	2–5 years ago	1
	>5 years ago	2

*Other historical variables, such as high myopia or hyperopia, systemic hypertension, corticosteroid use, and perhaps diabetes, are not strong enough to be assigned a weight but may be considered in the overall assessment of glaucoma risk.

Level of Glaucoma Risk	Weighting Score
High	4 or greater (referral advisable)
Moderate	3 (referral advisable)
Low	2 or less

Cataract is the most common cause of decreased vision (not correctable with glasses) in the United States. However, it is one of the most successfully treated conditions in all of surgery. Approximately 1.64 million cataract extractions are done each year in the United States, usually with implantation of an intraocular lens. If an implant is not used, visual rehabilitation is still possible with a contact lens or thick (aphakic) eyeglasses.

If cataract surgery is considered, it is important to be certain that visual loss is explained fully by cataract or significantly by the cataract if other ocular conditions such as glaucoma, macular degeneration, or diabetic retinopathy also exist. Assessment of the visual significance of the cataract is more difficult with coexisting ocular pathology. Sometimes cataract surgery is performed to facilitate diagnosis or treatment of other ocular diseases.

BASIC INFORMATION

This section provides an overview of the lens as well as cataracts and their symptoms.

LENS

The crystalline lens focuses a clear image on the retina. The lens is suspended by thin filamentous zonules from the ciliary body between the iris anteriorly and the vitreous humor posteriorly. Contraction of the ciliary muscle permits focusing of the lens. The lens is enclosed in a capsule of transparent elastic basement membrane. The capsule encloses the cortex and the nucleus of the lens as well as a single anterior layer of cuboidal epithelium. The lens has no innervation or blood supply. Nourishment comes from the aqueous fluid and the vitreous.

The normal lens continues to grow throughout life. The epithelial cells continue to produce new cortical lens fibers, yielding a slow increase in size, weight, and density over the years. The normal lens consists of 35% protein by mass. The percentage of insoluble protein increases as the lens ages and as a cataract develops.

CATARACT

A cataract is any opacity or discoloration of the lens, whether a small, local opacity or the complete loss of transparency. Clinically, the term *cataract* is usually reserved for opacities that affect visual acuity because many normal lenses have small, visually insignificant opacities.

A cataract is described in terms of the zones of the lens involved in the opacity. These zones of opacity may be subcapsular, cortical, or nuclear and may be anterior or posterior in location. In addition to opacification of the nucleus and cortex, there may be a yellow or amber color change to the lens. A cataract also can be described in terms of its stage of development. A cataract with a clear cortex remaining is immature. A mature cataract (Figure 3.9) has a totally opacified cortex.

The most common cause of cataract is age-related change. Other causative factors include trauma, inflammation, metabolic and nutritional defects, and the effects of corticosteroids. Cataracts may develop very slowly over the years or may progress rapidly, depending on the cause and type of cataract.

SYMPTOMS OF CATARACT

Patients may first notice image blur as the lens loses its ability to resolve separate and distinct objects. Patients are first aware of a disturbance of vision, then a diminution, and finally a failure of vision. The degree of visual disability caused by a cataract depends on the size and location of the opacity. Axial opacities—affecting the nucleus or central subcapsular areas—cause much more disabling visual loss than do peripheral opacities.

Patients with nuclear sclerosis may develop increasing lenticular (ie, referring to the crystalline lens) myopia because of the increased refractive power of the denser nucleus. As the size of the cataract increases, patients become

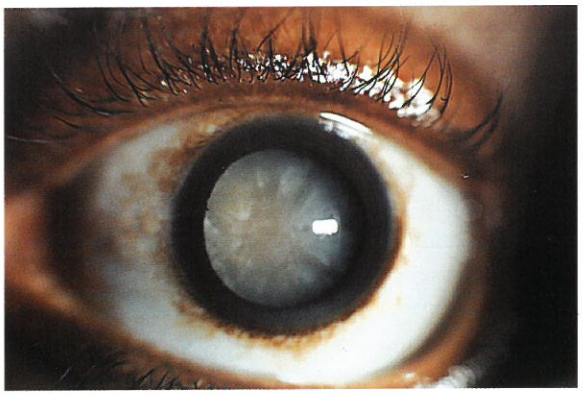


FIGURE 3.9 Mature cataract. A cataract is called *mature* when the lens is totally opacified. A red reflex cannot be obtained; the pupil appears white. The radial spokes in this figure reflect variations in density of the radially arranged fibers in the cortical layers of the lens. Light still reaching the retina is totally diffused and will allow the perception of light but not form.

progressively more myopic. Patients may find they can read without the glasses normally required, a phenomenon often called *second sight*. Patients may note monocular double or multiple images, due to irregular refraction within the lens. They frequently complain of "starbursts" around lights and difficulty with night driving. With yellowing of the lens nucleus, objects appear browner or yellower and color discrimination is more difficult.

Patients with posterior subcapsular cataracts (Figure 3.10) may note a relatively rapid decrease in vision, with glare as well as image blur and distortion. In contrast to nuclear cataracts (Figure 3.11), posterior subcapsular cataracts frequently affect the near vision. This type of cataract may be associated with metabolic causes such as diabetes mellitus or corticosteroid use.

Eventually all cataracts lead to a generalized impairment of vision. The degree of visual disability may vary depending on lighting and tasks the patient needs to perform.

WHEN TO EXAMINE

A patient with decreasing vision requires a complete examination to determine the cause of the visual decline. Evaluation of the visual significance of a cataract includes optic nerve and retinal evaluation to detect coexisting disease that could also affect the visual acuity. Special attention is given to the macula when a patient reports difficulty with near work or metamorphopsia (ie, a wavy distortion of central vision). In the presence of coexisting disease, the ophthalmologist may order special tests to better determine the contribution of the cataract to the decreased visual acuity.

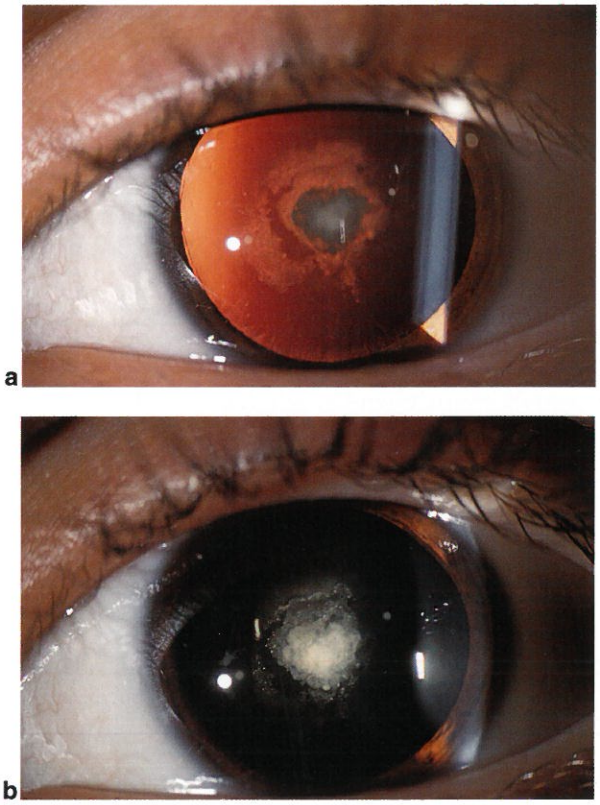


FIGURE 3.10 Posterior subcapsular cataract. **a.** The red reflex has a central dark shadow. **b.** Posterior subcapsular cataracts can be seen in younger patients and may be associated with chronic steroid use. *(Courtesy Cynthia A. Bradford, MD.)*

If the lens is densely cataractous, the ophthalmoscope will not provide a view of the fundus through the opacity. In this situation, retinal and optic nerve pathology cannot be adequately assessed. For example, if the patient has diabetic retinopathy in the contralateral eye there is increased risk for a portion of the visual loss in the eye with cataract to be related to diabetic retinopathy. Removal of the cataract would allow evaluation and possible treatment of the retinal pathology. The patient should understand the expected visual outcome of surgery.

HOW TO EXAMINE

In addition to obtaining an accurate history of visual decline, the following examination methods are particularly helpful in determining whether visual loss is attributable to cataract, to some other cause, or to a combination of causes:

- **Visual acuity** The first step in any evaluation of visual decrease is the measurement of visual acuity. Refer to Chapter 1 for details.
- **Pupillary responses** Chapter 7 describes how to perform a basic pupillary examination and provides details on the neurologic implications of pupillary responses. Even an advanced cataract would not produce a relative afferent pupillary defect.

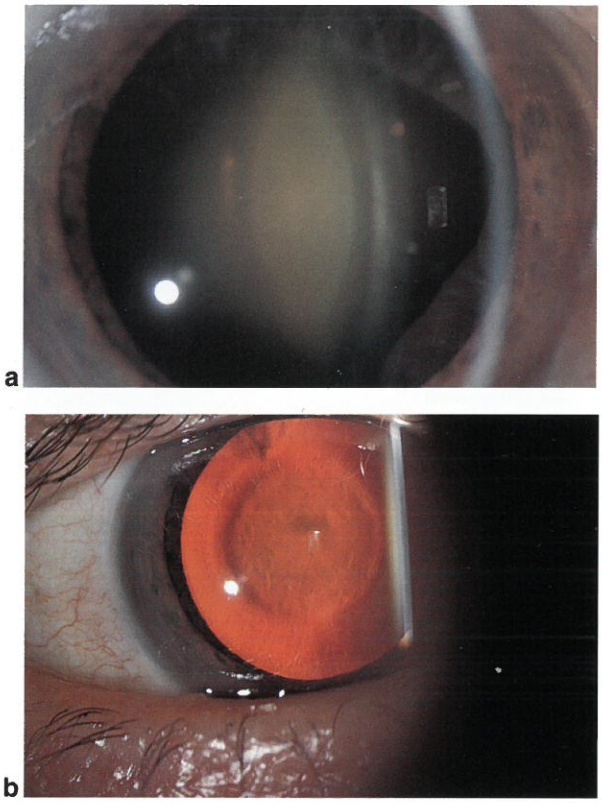


FIGURE 3.11 Nuclear cataract. **a.** Slit-lamp photograph of nuclear sclerotic cataract demonstrates the yellow coloration of the cataract and the central granular opacification. **b.** Red reflex photograph of the same cataract highlights the central distortion of the lens that results in the decreased visual acuity as well as the phenomenon of starbursts around lights. This can easily be seen with the direct ophthalmoscope if the pupil is dilated. *(Courtesy Cynthia A. Bradford, MD.)*

- **Ophthalmoscopy** The examiner's view into the eye should be about the same as the cataract patient's visual acuity; that is, the cataract should affect the physician's view into the eye through the direct ophthalmoscope to about the same extent as it does the patient's view out of the eye.

HOW TO INTERPRET THE FINDINGS

An early cataract is not visible to the unaided eye. If the cataract becomes very dense, it may appear as a white pupil, or leukocoria. The lens can be evaluated with the ophthalmoscope using a plus-lens setting. The lens opacification with a partial cataract will appear black against the red reflex of the fundus. Generally, the denser the cataract, the poorer the red reflex and the worse the visual acuity.

In addition to ophthalmoscopy, an ophthalmologist would routinely perform a slit-lamp examination, which provides a magnified, stereoscopic view of the lens and other anterior segment structures. A funduscopic examination is needed to evaluate the macula and optic nerve for disease that could be contributing to the visual loss.

MANAGEMENT OR REFERRAL

It is important not to assign visual loss to cataract before ensuring that other, more serious causes of visual loss have not been overlooked. The decision to

refer a patient with cataract should be based in part on whether or not the cataract keeps the patient from doing what he or she wants to do. A cataract can interfere with patients' daily activities of living by limiting their ability to drive safely, read, or participate in sports or other hobbies. Patients with cataract-associated visual loss that negatively affects their daily living may benefit from a surgical procedure of cataract extraction with intraocular lens implantation. A prospective study has shown that cataract extraction has been associated with a 50% drop in motor vehicle accidents. Uncorrected, chronic vision loss in older patients in residential care has been associated with an increased risk of falls and falling injuries. The Framingham Study found an increase in the relative risk of hip fracture with moderate (20/30 to 20/80) and severe (20/100 or worse) vision impairment over 10 years of 1.54 and 2.17, respectively. Even moderate vision loss in one eye alone was associated with an increased risk of 1.94, suggesting the role of good stereoscopic vision in prevention of falls and fractures.

After cataract-removal surgery, many patients require a laser surgical procedure to open an opacified posterior capsule. This has led to a popular misconception that a cataract can actually be removed with a laser. The operating surgeon should preoperatively examine all patients who are contemplating cataract surgery. A discussion with the surgeon of the risks, benefits, and alternatives of the surgery, as well as planning for postoperative care, is imperative.

MACULAR DEGENERATION

RELEVANCE

In the United States, age-related macular degeneration is the leading cause of irreversible central visual loss (20/200 or worse) among people age 50 or older. Because certain types of macular degeneration can be effectively treated, it is important to recognize this entity and to refer for appropriate care. It is important to distinguish between the possible causes of visual loss, whether cataract (surgically correctable), glaucoma (medically or surgically treatable), or macular degeneration (potentially treatable medically or by laser surgery).

BASIC INFORMATION

This section introduces macular anatomy and macular changes due to aging.

MACULAR ANATOMY

The macula is situated between the temporal vascular arcades. The center of the macula, the fovea, is an oval area situated about 2 disc diameters temporal

and slightly inferior to the optic disc (see Figure 1.15 for a depiction of the normal fundus). The macula is composed of both rods and cones and is the area responsible for detailed, fine central vision. The fovea (Figure 3.12) is partly avascular and appears darker than the surrounding retina. The foveola is the pit-like depression in the center of the macula. Here, there is a high density of cones but no rods are present. The central depression of the fovea may act like a concave mirror during ophthalmoscopy, producing a light reflection (ie, foveal reflex).

AGE-RELATED MACULAR CHANGES

Macular changes due to age include drusen, degenerative changes in the retinal pigment epithelium, and choroidal neovascular membranes.

Drusen are hyaline nodules (or colloid bodies) deposited in Bruch's membrane, which separates the retinal pigment epithelium (the outermost layer of the retina) from the inner choroidal vessels. Drusen may be small and discrete (Figure 3.13) or larger, with irregular shapes and indistinct edges. Patients with drusen alone tend to have normal or near-normal visual acuity, with minimal metamorphopsia. Drusen may be seen with increasing age, with retinal or choroidal degeneration, and as a primary dystrophy.

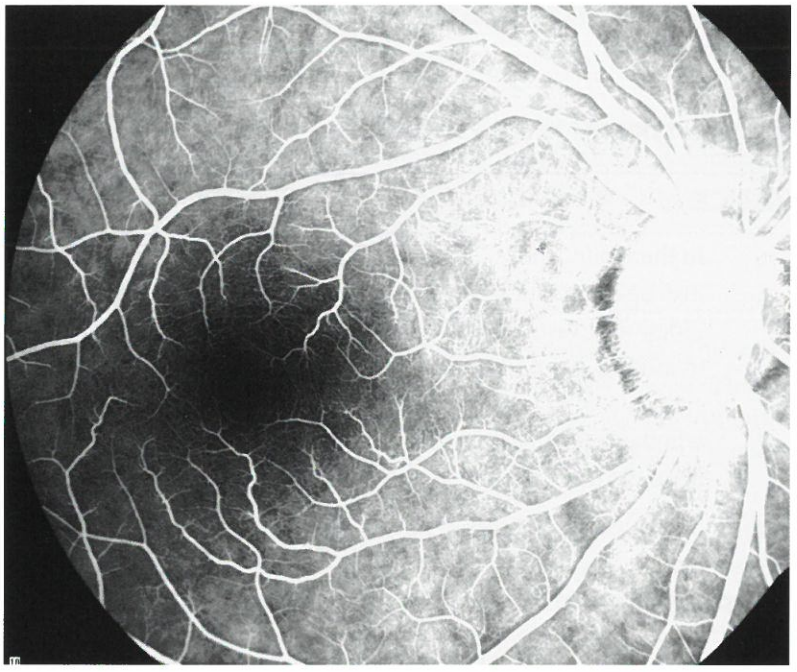


FIGURE 3.12 Fovea. The fovea is avascular, as demonstrated in this fundus fluorescein angiogram. The central capillary-free zone identifies the foveal region. *(Courtesy Reagan Bradford, MD.)*

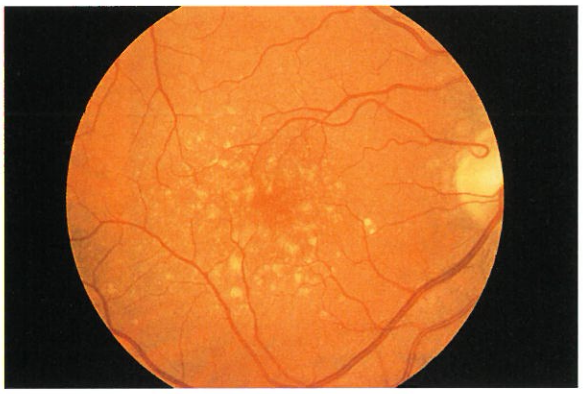


FIGURE 3.13 Drusen. Small and medium-sized drusen (yellow deposits) beneath the right macular in this patient with nonexudative "dry" age-related macular degeneration. Although acuity may be normal initially, these lesions can lead to significant visual loss if the central macula becomes involved. *(Courtesy Ronald M. Kingsley, MD.)*

Degenerative changes in the retinal pigment epithelium itself may occur with or without drusen. These degenerative changes are manifested as clumps of hyperpigmentation or depigmented atrophic areas (Figure 3.14). The effect on visual acuity is variable.

About 20% of eyes with age-related macular degeneration develop choroidal neovascularization, producing the so-called *neovascular*, or "wet," macular degeneration. The extension of vessels from the inner choroid layer into

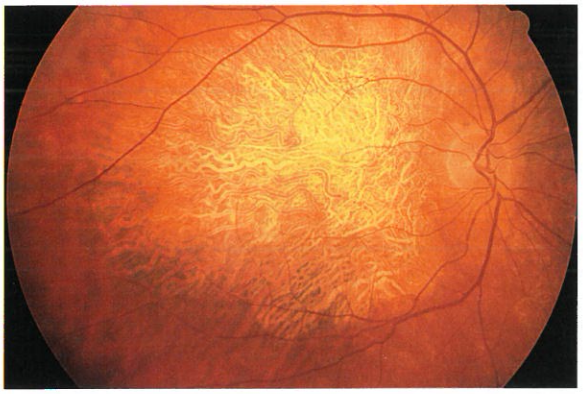


FIGURE 3.14 Retinal pigment epithelial atrophy. An extensive area of geographic, submacular pigment atrophy involves the entire posterior retina between the temporal retinal vascular arcades. The underlying choroidal vasculature is more prominent when the pigment epithelium is absent or atrophic. This patient demonstrates advanced nonexudative, or "dry," age-related macular degeneration. Such a patient typically has poor visual acuity. *(Courtesy Ronald M. Kingsley, MD.)*

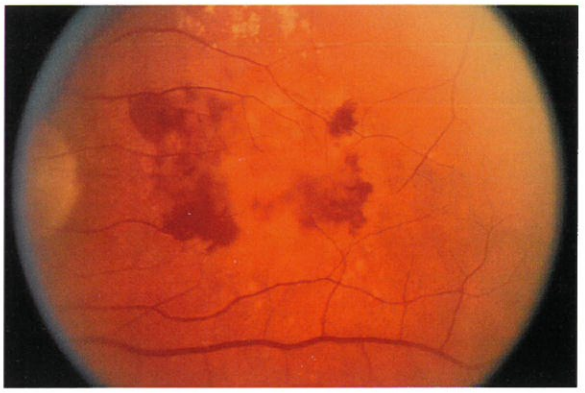


FIGURE 3.15 Subretinal hemorrhage. The subfoveal fluid, hemorrhage, and exudate present in this patient indicate choroidal neovascularization. These findings are often present in patients with neovascular, or "wet," age-related macular degeneration. *(Courtesy Ronald M. Kingsley, MD.)*

the subpigment epithelial space and eventually into the subretinal space occurs through a defect in Bruch's membrane.

The choroidal neovascular net may be associated with subretinal hemorrhage and exudates, and with fibrosis in the late or disciform stage (Figure 3.15). Subretinal hemorrhage may result in acute visual loss (see Chapter 2). The larger the membrane and the closer to the center of the fovea, the worse the prognosis for good central vision.

Fluorescein angiography, a technique utilized by ophthalmologists, may be necessary to identify neovascularization and is mandatory before considering laser surgery. Intravenous injection of fluorescein dye and subsequent rapid-sequence photography help demonstrate the retinal and choroidal vasculature. In contrast to competent retinal veins and arteries, new vessels can be identified because they leak fluorescein dye. In addition, because the retinal pigment epithelium normally acts as a physical and optical barrier to fluorescein, angiography facilitates identification of pigment epithelial defects. Indocyanine green is another dye used to demonstrate choroidal neovascularization.

Compare Figure 3.15, a fundus photograph depicting a subretinal hemorrhage implying the presence of choroidal neovascularization, with Figure 3.16,

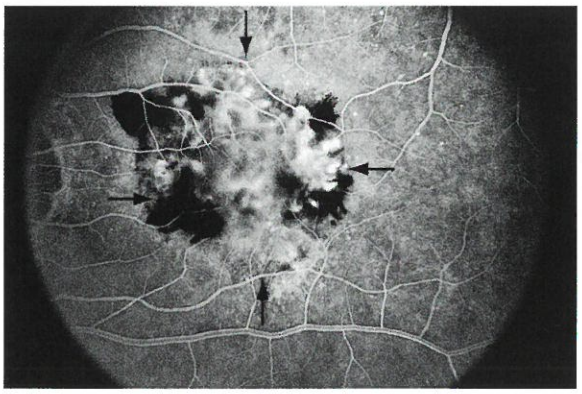


FIGURE 3.16 Neovascular net. The fluorescein frame depicts the neovascular net in the fundus photograph in Figure 3.15. The hyperfluorescent (bright) area under the left fovea highlights the area of neovascularization and is used to determine the management strategy. *(Courtesy Ronald M. Kingsley, MD.)*

a fundus fluorescein angiogram of the same eye, which reveals the choroidal neovascularization and associated subretinal hemorrhage.

Age-related changes are typically confined to the posterior pole of the eye. Thus, the patient with macular degeneration may have very poor central vision, but will tend to retain functional peripheral vision. Visual aids, such as high-plus magnifiers and telescopic devices, may help the patient. In addition to age, other causes of chronic maculopathy include heredity and metabolic changes.

WHEN TO EXAMINE

Any patient with decreasing vision requires examination to determine the cause of the visual change. In assessing a patient with blurred or distorted central vision, every effort should be made to examine the macula with the ophthalmoscope. Of course, opacities in the cornea, lens, or vitreous may preclude an adequate view of the macula.

HOW TO EXAMINE

The following techniques are especially helpful in evaluating macular degeneration as the cause of visual decrease or major changes in central vision:

- **Visual acuity measurement** Refer to Chapter 1 for instructions.
- **Amsler grid testing** Amsler grid testing (Figure 3.17) is a useful method of evaluating the function of the macula. The test is carried out by having

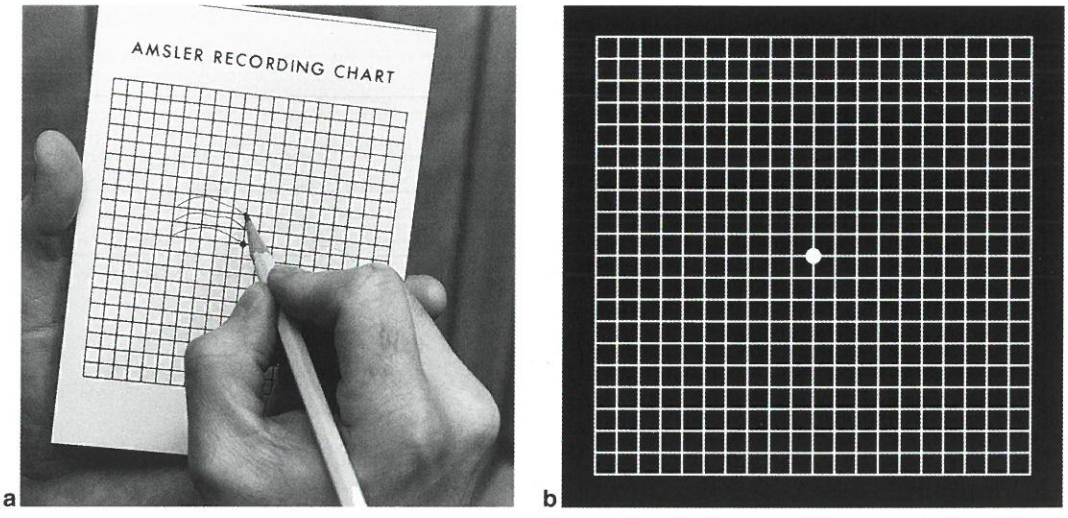


FIGURE 3.17 Amsler grid testing. **a.** The patient indicates the nature and location of his central field defect by sketching what he perceives on the Amsler grid. **b.** The typical grid pattern of white lines on a black background.

the patient look with one eye at a time at a central spot on a page where horizontal and vertical parallel lines make up a square grid pattern. This grid pattern may be printed with white lines against a black background or vice versa. The patient is asked to note irregularities in the lines. Irregularities may be reported as lines that are wavy, seem to bow or bend, appear gray or fuzzy, or are absent in certain areas of the grid, indicating a scotoma. The straight line, right angle, and square are geometric shapes that highlight areas of distortions most easily. With the chart held at a normal reading distance of 30 cm from the eye, the Amsler grid tests the central 20 degrees of visual field. Thus, the entire macula is evaluated with this examination.

- **Ophthalmoscopy** The macular area is studied with the direct ophthalmoscope. Sometimes it is helpful to have the patient look directly into the light of the instrument. Dilation of the pupil is usually necessary for adequate examination.
- **Additional studies** The ophthalmologist may elect to carry out special studies to better evaluate the macula and macular function. Procedures such as stereoscopic slit-lamp examination and fluorescein angiography may be necessary to determine pathologic changes.

HOW TO INTERPRET THE FINDINGS

The appearance of the macula often does not accurately predict the visual acuity. The macula may look more or less involved than the vision indicates. Drusen, areas of decreased or increased pigmentation, subretinal exudate, and hemorrhage are all important signs to check for in a direct ophthalmoscopic examination of the macula. The absence of the foveal reflex and a mottled appearance of the underlying retinal pigment epithelium are among the early signs of macular disease.

MANAGEMENT OR REFERRAL

Any patient who has one or more of the following should be referred to an ophthalmologist:

- Recent onset of decreased visual acuity
- Recent onset of metamorphopsia (central visual distortion)
- Recent onset of a scotoma (blind spot)
- Any ophthalmoscopic abnormalities in the appearance of the macula, such as drusen, degenerative changes in the retinal pigment epithelium, exudate, or blood

Although a patient with metamorphopsia may have drusen in the macula only, many patients with this complaint have complicating choroidal neovas-

cularization and most need fluorescein angiography to establish their management. Clinical studies have indicated that argon laser photocoagulation of choroidal neovascular membranes that are not too close to the fovea significantly reduces the risk of central visual loss. When patients have choroidal neovascularization that extends beneath the fovea, they may be a candidate for photodynamic therapy with verteporfin (Visudyne). Verteporfin is given intravenously and selectively adheres to the neovascular membrane. A cold (nonthermal) laser is then used to activate the verteporfin treatment effect by reducing the growth of and leakage from the neovascular membrane. Visudyne therapy has been proven effective in reducing the risk of visual loss in this situation.

A number of additional treatments for neovascular AMD are being evaluated in clinical trials. Transpupillary thermotherapy (TTT), subretinal surgery (SST), rheopheresis, and anti-angiogenic agents that are injected into or around the eye have generated hope for the future therapy of "wet" AMD. Some patients undergo therapies, such as microcurrent stimulation and acupuncture, that have yet to undergo rigorous clinical trial. Primary care physicians may be asked their opinion on these therapies.

It is possible that neovascular AMD will eventually be best treated by a combination of therapies. Because many of the available treatments are costly, the patient with neovascular AMD needs to carefully assess the existing evidence for treatment effectiveness before proceeding.

Current therapies for neovascular AMD offer hope for patients with acute symptoms of visual loss (usually less than 3 months' duration). After several months, it is unlikely that treatment will benefit the patient visually.

There is ongoing research on the effect of vitamins on the development and progression of AMD. Patients should be encouraged to eat a diet rich in green (kale, collard greens, spinach, broccoli) and yellow vegetables as well as take a daily multivitamin. They should also be cautioned that smoking is associated with more severe AMD.

In addition, patients who are 50 years of age or older with large drusen should be evaluated by an ophthalmologist to consider their need for oral supplementation of a daily dose of antioxidants (500 mg of vitamin C, 400 IU of vitamin E, and 15 mg of beta carotene) and minerals (80 mg of zinc oxide and 2 mg of cupric oxide) to reduce the risk of vision loss from neovascular macular degeneration or geographic atrophy. This supplementation is modified for smokers to exclude beta carotene because it may increase their risk of developing lung cancer.

THE VISUALLY IMPAIRED PATIENT

Despite medical or surgical therapy, some patients will have a significant residual visual impairment. These patients are candidates for low vision services