

*Care of the Patient with*  
**Age-Related  
Macular  
Degeneration**



**OPTOMETRY:  
THE PRIMARY EYE CARE PROFESSION**

Doctors of optometry are independent primary health care providers who examine, diagnose, treat, and manage diseases and disorders of the visual system, the eye, and associated structures as well as diagnose related systemic conditions.

Optometrists provide more than two-thirds of the primary eye care services in the United States. They are more widely distributed geographically than other eye care providers and are readily accessible for the delivery of eye and vision care services. There are approximately 32,000 full-time equivalent doctors of optometry currently in practice in the United States. Optometrists practice in more than 7,000 communities across the United States, serving as the sole primary eye care provider in more than 4,300 communities.

The mission of the profession of optometry is to fulfill the vision and eye care needs of the public through clinical care, research, and education, all of which enhance the quality of life.



# OPTOMETRIC CLINICAL PRACTICE GUIDELINE

## CARE OF THE PATIENT WITH AGE-RELATED MACULAR DEGENERATION

### Reference Guide for Clinicians

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NOTE: Clinicians should not rely on the Clinical  
Guideline alone for patient care and management.  
Refer to the listed references and other sources  
for a more detailed analysis and discussion of  
research and patient care information. The  
information in the Guideline is current as of the  
date of publication. It will be reviewed periodically  
and revised as needed.

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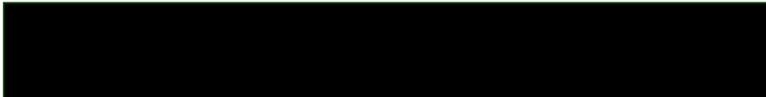


## **INTRODUCTION**

Optometrists, through their clinical education, training, experience, and broad geographic distribution, have the means to provide effective primary eye and vision care for a significant portion of the American public, including older Americans. Age is a risk factor for many ocular and visual disorders. Increased accessibility to care may reduce the risk of significant vision loss for certain individuals.

This Optometric Clinical Practice Guideline for Care of the Patient with Age-Related Macular Degeneration (AMD) describes appropriate examination and treatment procedures to help reduce severe vision loss from age-related macular degeneration by identifying patients with high-risk characteristics. It contains recommendations for examination, treatment, and when necessary, referral for consultation with or treatment by another health care provider. This Guideline will assist optometrists in achieving the following goals:

- Identify ocular, personal, and environmental risk characteristics for AMD
- Accurately diagnose AMD
- Develop a decision making strategy for management of patients at risk for severe vision loss from AMD
- Provide information and resources for appropriate patient education in the area of vision rehabilitation
- Propose a philosophy and rationale for management and prevention of AMD.



## **I. STATEMENT OF THE PROBLEM**

Age-related macular degeneration is an acquired retinal disorder with far-reaching psychosocial and economic implications. As the leading cause of legal blindness (visual acuity of 20/200 or worse) for persons over age 65 in the United States, it accounts for 14 percent of new legal blindness, with 16,000 cases reported annually.<sup>1-3</sup> AMD is the leading cause of severe vision loss in persons over age 50 and it is second only to diabetes as the leading cause of blindness in the 45 to 64 year-old age group.<sup>4-6</sup> "Severe vision loss" is categorized as visual acuity of 20/200 or worse. "Significant vision loss" refers to a loss of visual function that interferes with customary or required activities or lifestyle, usually at a level approximating 20/50-20/70 or worse.

Although AMD is not curable, in some cases severe vision loss can be prevented because certain forms of the disease respond favorably to laser treatment, especially with early diagnosis and prompt intervention. Certain preventive measures, including appropriate long-term surveillance, patient education, lifestyle changes, and careful evaluation, can reduce ocular morbidity. Only a small percentage of patients with AMD will benefit from laser photocoagulation treatment; identifying candidates for treatment is critical in attempting to prevent severe vision loss.

The number of Americans over age 65 will more than double between the years 1990 and 2020.<sup>7</sup> Because age is a significant risk factor for the development of AMD, timely access to eye care may have preventive value. Many older Americans neither seek nor have access to regular eye care; thus the risk for vision loss in this population is unnecessarily high if AMD is not diagnosed promptly.

Eighty percent of the anticipated 2 million Americans who will be residing in nursing facilities by the year 2000 will be over age 75. The number of Americans needing long-term care is projected to increase from 4 million to 18 million by the year 2040.<sup>8</sup> Their access to care may be limited in certain settings, especially extended care facilities. Without timely diagnosis and treatment, loss of vision in these environments cannot be prevented. The onset of AMD is insidious. Coupled with

environmental and lifestyle factors which may play secondary, but important, roles in the development of the disease, the nature of AMD makes patient education, early detection, and referral critical for high-risk patients.

The Macular Photocoagulation Study (MPS) initiated in 1979 investigated the efficacy of laser photocoagulation for treatment of AMD.<sup>9</sup> This prospective study to evaluate the risk for severe vision loss from AMD demonstrated that some patients with the exudative form of the disease benefit from laser photocoagulation when AMD is identified and treated early. Comprehensive eye care is of increased importance because of the high recurrence rate of choroidal neovascular membranes and the rapid loss of vision through the exudative process in AMD.

### **A. Description and Classification of Age-Related Macular Degeneration**

Age-related macular degeneration is an acquired retinal disorder which is characterized by any of the following fundus changes: pigmentary atrophy and degeneration, drusen and lipofuscin deposits, and exudative elevation of the outer retinal complex in the macular area. AMD, which usually occurs in patients over age 55, results in progressive, sometimes significant, irreversible loss of central visual function from either fibrous scarring or diffuse, geographic atrophy of the macula. The definition can be expanded to include extrafoveal lesions that would have an impact on vision if superimposed on the foveal region.<sup>10</sup> The ICD-9-CM classification of AMD is contained in Appendix Figure 3.

#### **1. Nonexudative AMD**

Nonexudative (dry or atrophic) AMD accounts for 90 percent of all patients with AMD in the United States.<sup>2</sup> The disorder results from a gradual breakdown of the retinal pigment epithelium (RPE), the accumulation of drusen deposits, and loss of function of the overlying photoreceptors. Most patients with nonexudative AMD experience gradual, progressive loss of central visual function. This loss of vision is more noticeable during near tasks, especially in the early stages of the disease. In an estimated 12-21 percent of patients, nonexudative AMD

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progresses to cause vision levels of 20/200 or worse.<sup>3,6,11</sup> Both choroidal neovascularization (CNV) and subretinal or sub-RPE exudation are conspicuously absent in this category of AMD.

#### **2. Exudative AMD**

Although exudative (wet) AMD accounts for only 10 percent of patients with AMD, 90 percent of the AMD patients with significant vision loss have this form of the disease.<sup>6,12,13</sup> Exudative AMD is characterized by the development of neovascularization in the choroid, leading to serous or hemorrhagic leakage and subsequent elevation of the RPE or neurosensory retina. Patients with exudative AMD tend to notice a more profound and rapid decrease in central visual function. Serous or hemorrhagic leakage from the new choroidal vessels causes dysmorphopsia, scotoma, and blurred vision.<sup>14</sup>

Each of the two forms of AMD has its own features. In most patients nonexudative AMD will not progress to severe vision loss. Those patients in whom AMD progresses to the exudative form are at greatest risk for severe visual impairment. Patients who have exudative maculopathy with drusen in the fellow eye are at significant risk of developing CNV.<sup>15</sup>

#### **3. Geographic Atrophy**

Geographic atrophy is a clinical manifestation of progressive atrophy of the RPE in conjunction with drusen formation. In most cases the natural history of the disease reflects retinal atrophy following the fading of drusen.<sup>16</sup> From one to several well-demarcated areas of RPE atrophy are accompanied by overlying photoreceptor damage. The process is diffuse, often bilateral and symmetrical, and it can lead to vision loss even in the absence of choroidal neovascularization. CNV can develop as a separate entity in the presence of soft, confluent drusen. Geographic atrophy can also follow the collapse of a retinal pigment epithelial detachment though it is uncommon, occurring in less than 2 percent of serous RPE detachment cases within 2 years of diagnosis.<sup>17-20</sup> Sometimes geographic atrophy occurs after an RPE tear or rip and can be

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associated with ill-defined or occult choroidal neovascular membranes (CNVM).

#### **4. Stages of AMD**

Numerous systems have been proposed for classifying the various stages of AMD (Table 1). Most of these systems rely on the ophthalmoscopic appearance of the individual macular lesions, the extent of involvement of the macula, and the patient's visual acuity.<sup>21</sup> The early stage of AMD is recognizable by macular changes consisting of RPE abnormalities and typical, soft, and indistinct drusen. Patients who have fundus findings in this category can be considered at risk for severe vision loss especially if CNV develops later. Late or advanced stages of AMD are characterized by the presence of CNV with exudation, consisting of blood, lipid, or serosanguinous material, that produces RPE or neurosensory detachment. The final stage of the exudative form of AMD consists of a disciform scar in the foveal area with severe loss of central visual function.

**Table 1**  
**Stages of AMD \*\***

1. Pigmentary abnormalities:
  - RPE degeneration
  - Increased retinal pigment in the macular area
2. Retinal pigment epithelial degeneration and increased retinal pigment:
  - Granules or clumps of gray or black pigment in or beneath the retina
  - Grayish-yellow or pinkish-yellow areas of varying density and configuration in the plane of the RPE
3. Early AMD:
  - Soft, indistinct, or reticular drusen
  - Any type of drusen (except hard, indistinct) with RPE degeneration in the absence of signs of late AMD
4. Late AMD:
  - Exudative AMD
    - RPE or serous detachment of the sensory retina
    - Subretinal or pigment epithelium hemorrhage
  - Pure geographic atrophy
5. Pure geographic atrophy:
  - Geographic atrophy in the absence of exudative macular degeneration
  - Loss of RPE and increased visualization of the underlying choroid

---

\* The classification of the stages of AMD is not clearly defined; rather, the various categories have been delineated based on ophthalmoscopically visible lesions. The transition from one stage or class to another is not always obvious or clearly identifiable. Retinal drusen are categorized according to size, confluence (touching or merging), uniformity of appearance, and sharpness (soft, indistinct or distinct; hard, indistinct or distinct; or reticular).

## **B. Epidemiology of Age-Related Macular Degeneration**

### **1. Prevalence and Incidence**

Age-related macular degeneration is the second leading cause of legal blindness in the United States and other developed countries<sup>1,2,12,22</sup> and is the leading cause in persons over age 65.<sup>23</sup> Almost three-quarters of a million Americans have visual acuities of 20/200 or worse in one or both eyes as a result of AMD. Ninety percent of them are a result of the exudative form of the disease. AMD accounts for 16,000 cases or 14 percent of new legal blindness yearly.<sup>24-26</sup> Among patients over the age of 52, 9 percent suffer from one or more forms of AMD; and 6.4 percent of those between the ages of 65 and 74 have AMD in one or both eyes.<sup>12,27,28</sup> The prevalence of AMD and severe vision loss increases with age, and approximately 20-30 percent of persons over age 75 are affected. Applying standard diagnostic criteria, one-third of males and one-fourth of females over age 75 have some form of AMD.

### **2. Risk Factors**

Numerous factors are implicated in the development and progression of ocular changes that result in AMD. Factors which should be identified and considered in the examination, diagnosis, and management of the patient include age, gender, race, and ocular, hereditary, systemic, and environmental factors.

#### **a. Age**

Although age is a significant risk factor, increased longevity does not predestine one to develop AMD. However, it is likely that the number of persons with AMD will increase dramatically as the number of persons over age 65 in the United States increases during the next 30 years. Because exudative AMD begins at a later age, increased longevity may result in a higher incidence of vision loss from this disease.

**b. Gender**

Early studies showed that females have a higher incidence of AMD than males but this may have been attributed to greater life expectancy.<sup>1</sup> Females also demonstrate an earlier age of onset of AMD.<sup>29</sup>

**c. Race**

The incidence of AMD is higher in Caucasians than African Americans and Asians, among whom it has been reported to be rare.<sup>30</sup>

**d. Ocular Factors**

Hyperopia greater than 0.75 diopters (D) increases the risk of exudative AMD by up to 2.5 times.<sup>6,31,32</sup> In some studies light ocular pigmentation has been identified as a risk factor for both forms of AMD although pigmentation does not seem to be an important factor for neovascular AMD according to some reports.<sup>31,33</sup>

**e. Hereditary Factors**

Some genetic predisposition for AMD seems to exist and 10-20 percent of patients with AMD have at least one first-degree family member with vision loss.<sup>6,25</sup> Studies have reported AMD with vision loss in at least one parent or sibling of affected patients.

**f. Systemic Factors**

Cardiovascular disease, including circulatory problems, arteriolar sclerosis, stroke, heart attack, and angina, has been implicated as a secondary risk factor in patients with AMD.<sup>6,28,34-37</sup> The association between AMD and elevated serum cholesterol levels is questionable.<sup>32</sup> No direct relationship, other than its association with cardiovascular disease, has been found between hypertension and AMD.<sup>6,34</sup>

Evidence of a relationship between cigarette smoking and an increased risk of AMD has been reported in several studies.<sup>38-40</sup> Current smokers are 2.4-2.5 times more likely to develop AMD than individuals who

never smoked. Cigarette smoking has been proposed to affect choroidal circulation, eventually leading to the loss of RPE. This appears to be particularly evident in persons with exudative AMD, especially in males currently smoking, and CNV appears to have a higher rate of recurrence (1.7-2.2 times) in current smokers.<sup>36</sup>

**g. Environmental Factors**

Cumulative light toxicity may damage the outer retina, contributing to the changes that lead to the development of AMD. The influence of antioxidants that protect against damage to the RPE and photoreceptors requires further evaluation.<sup>41</sup>

**C. Clinical Background of Age-Related Macular Degeneration**

**1. Natural History**

Improved awareness of the etiopathogenesis and clinical course of AMD has led to better diagnosis and treatment strategies. Most patients with severe vision loss have exudative AMD. However, patients with atrophic AMD may experience a gradual but significant loss of visual function or may develop CNV and progress to the exudative stage of the disease. Therefore, both types of patients must be monitored closely. Specific retinal lesions will predispose the patient to significant vision loss. Signs of exudative AMD (including a subretinal greenish-gray lesion, serous elevation of the RPE or neurosensory retina, intraretinal hemorrhage, and exudate) warrant close surveillance or immediate referral of the patient to a retina specialist, if available, or a general ophthalmologist experienced in retinal disease for fluorescein angiography (FA).

Preliminary data from the Macular Photocoagulation Study demonstrated that laser photocoagulation can prevent severe vision loss in a small percentage of persons with exudative maculopathy. In patients with well-defined CNVMs located from 200-2500 microns from the center of the foveal avascular zone (FAZ), treatment could be beneficial.<sup>9</sup>

## **2. Common Signs, Symptoms, and Complications**

Whereas AMD can represent accelerated or advanced stages of aging changes in the retina, a clear understanding of the natural history of AMD is important. Symptoms may not become apparent until late in the disease process, at which time treatment may not be effective. Selective patient identification on the basis of clinical signs and symptoms can help ensure appropriate and timely treatment. Other strategies, including surveillance and patient education, are also important in patient management. Patients within the following groups at risk should be screened for signs and symptoms of AMD:

- Persons over 60 years of age
- Persons with hypertension or cardiovascular disease
- Cigarette smokers
- Persons with a first-degree family (sibling or maternal) history of vision loss from AMD regardless of age
- Persons with aphakia or pre-1984 pseudophakia
- Persons whose history indicates significant cumulative light exposure.

The spectrum of clinical signs for AMD consists of focal and diffuse changes in the outer retinal complex. They occur most often in the foveal area and include retinal pigment abnormalities, drusen, geographic atrophy, choroidal neovascularization, and related complications such as RPE detachment and vitreous hemorrhage.

### **a. Retinal Pigment Abnormalities**

Retinal pigment abnormalities are considered the earliest retinal manifestations of AMD and consist of increased retinal pigmentary degeneration and atrophy in the plane of the RPE. A grayish-yellow or pinkish-yellow area in the macula is often surrounded by a halo of gray or black pigment clumps in or beneath the retina. Increased lipofuscin in the RPE and the accumulation of debris on and within Bruch's membrane result in loss of photoreceptor function.

### **b. Drusen**

Drusen are yellow to yellowish-white nodular deposits found in the deeper layers of the retina. Along with pigmentary abnormalities, drusen are often the earliest ophthalmoscopic signs of aging in the retina. Visual acuity may be normal at this stage. Drusen alone are not enough to satisfy the definition of AMD when vision is normal. Several types of drusen have been described. The lesions are categorized by size, confluence, uniformity, and sharpness of borders.<sup>15,25</sup> (Table 1) Some form of drusen are found in the macular area in 50-95 percent of persons over age 70. Among persons with drusen, 10-15 percent may eventually develop exudative manifestations of AMD.<sup>10,34,35</sup> Twenty-five percent of patients over age 60 have ophthalmoscopically visible drusen, and almost 100 percent demonstrate some form of drusen histologically.<sup>42</sup>

Eyes with drusen and exudation in the fellow eye are at higher risk of developing CNV.<sup>15,43</sup> Usually occurring in clusters, drusen can be found anywhere in the posterior pole, but are most frequently found in the temporal parafoveal region.<sup>44</sup> Drusen can occur outside the vascular arcades and are often found on the nasal side of the optic disc. When found in the equatorial region, they are often accompanied by a reticular pattern of pigmented lines. There is an association between the diffuse drusenoid process encountered in the equatorial retina and geographic atrophy found in AMD.<sup>45</sup> Soft drusen are confluent lesions greater than 63 microns in diameter, that merge and touch and lack sharp borders.<sup>25</sup> Because these lesions may become vascularized, they represent a high risk for the development of CNVM. Pigmentary abnormalities are consistently found with both types of drusen. The terms "semisolid" or "mixed" drusen have been used to describe the clinical picture comprising both types of lesions.

### **c. Geographic Atrophy**

In nonexudative AMD geographic atrophy evolves from pigmentary degeneration and drusen formation in the macula. It is the most common cause for decreased visual function in AMD.<sup>46</sup> The atrophy is characterized by photoreceptor damage from loss of integrity of the underlying RPE. Drusen are present but do not appear to play an

important role in this process, although they do represent anatomic markers for RPE degeneration. Single or multiple areas of well-defined atrophy spread throughout the foveal and parafoveal area, producing a gradual decrease in vision. Geographic atrophy, which can also occur after the fading of drusen<sup>25</sup> or the collapse of an RPE detachment,<sup>17-20</sup> accounts for only 12-21 percent of severe vision loss in AMD.

**d. Choroidal Neovascularization**

Diffuse thickening of Bruch's membrane, in conjunction with soft, confluent drusen and pigment abnormalities, predisposes the patient to the development of a choroidal neovascular membrane. The new vessels of the CNVM form an organized yet fragile vascular system. As the system matures, the delicate neovascular twigs leak fluid (protein and lipids) into the subretinal, intraretinal, or sub-RPE space. Depending on numerous factors, the hemorrhage at the site of the membrane or in the subretinal space may extend into the vitreous.

In the case of sub-RPE hemorrhage, direct examination of the macula reveals a discrete round to oval elevated lesion. Often surrounded by a subretinal hemorrhage, the lesion appears greenish-gray or dirty brown. A halo of pigment may also surround the lesion. Other findings include subretinal lipid or blood, associated with an overlying thickened, detached neurosensory retina. This insidious lesion, which tends to be rapid growing and recurrent, heralds the onset of late AMD and exudative maculopathy. Numerous other ocular disorders involve CNV. Especially in persons under age 50, CNV must be differentiated from that found in AMD (Table 2). Age-related macular degeneration is the most common cause of CNV. When found with AMD, the lesions tend to be larger, probably because they are part of a more diffuse retinal disease process. These lesions are often found under the center of the FAZ which explains their more devastating effect on central visual function.<sup>47,48</sup> CNV, when associated with AMD, seems to induce more damage to the retinal tissue than that caused by other choroidal diseases, such as presumed ocular histoplasmosis syndrome, angioid streaks, or idiopathic causes of CNV.<sup>10</sup>

**Table 2**  
**Ocular Conditions with Choroidal Neovascular Membranes**

- Age-related macular degeneration (AMD)
- Presumed ocular histoplasmosis syndrome (POHS)
- Idiopathic choroidal neovascularization (CNV)
- Angioid streaks
- Myopic degeneration (lacquer cracks)
- Choroidal tumors
- Chronic uveitis
- Traumatic choroidal rupture
- Post-laser photocoagulation
- Toxoplasmosis.

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Choroidal neovascularization has been divided into two categories on the basis of clinical and fundus fluorescein angiographic appearance. Well-defined CNVs are delineated in their entirety during the early stages of the fluorescein angiogram. A further designation refers to those lesions which may not be entirely visible in the early stages of the fluorescein transit; however, the entire lesion is visible in the later stages following leakage. Classic, well-defined CNV, which fluoresces early in the angiogram, has a lacy appearance, with the cartwheel appearance of the new vessels sometimes apparent. The neurosensory retina has a hyperfluorescent pool of fluid in the late stages of the angiogram. Poorly defined membranes are difficult to identify due to the presence of subretinal blood, turbid fluid, or pigment. Half of the membranes are ill-defined and located within 200 microns of the center of the FAZ.<sup>49</sup>

A number of retinal pigment epithelial lesions occur in conjunction with CNV or occur as a consequence of the new vessel formation. A serous retinal pigment epithelial detachment (PED) resulting from leakage of new choroidal vessels is characterized ophthalmoscopically as a well-demarcated, dome-shaped, elevated yellowish-orange lesion. The overlying neurosensory retina may be detached, and the separation often exceeds the boundaries of the PED, providing further evidence of CNV. Hemorrhagic RPE detachment appears as a red or dark green elevated

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lesion. It may affect vision if located under the fovea. A hemorrhagic PED can mimic a choroidal melanoma and indicates the possible presence of a choroidal tumor with subretinal hemorrhage. Chorioretinal folds adjacent to the PED are sometimes observed. Long-standing detachments may lead to RPE atrophy or hypopigmentation in the area of the lesion. Patients over age 55 with RPE detachment and accompanying drusen are at significant risk of developing severe vision loss from CNV.<sup>17,19,20</sup>

Retinal pigment epithelial tears or rips occur spontaneously or follow laser photocoagulation of CNV. These RPE tears are a more consistent finding with occult or ill-defined CNV, usually occurring adjacent to the site of new choroidal vessels. Occasionally, RPE tears are detected with disciform scars and usually result in severe vision loss.<sup>50</sup>

Vitreous hemorrhage can occur with exudative AMD. When the patient reports sudden vision loss, it is sometimes the result of a breakthrough hemorrhage. Although the vitreous hemorrhage clears in 75 percent of patients, ultrasonography and stereoscopic fundus examination are indicated to rule out causes other than AMD.<sup>51</sup>

A fibrovascular disciform scar represents the final stage of untreated CNV. A yellowish-white to brown or black lesion is observed in the macula. Subretinal fluid or fresh hemorrhage frequently appears at the edges of the scar. Hypertrophic RPE, chorioretinal folds, and anastomosis of the retinal and choroidal circulations also have been observed in advanced CNV.

Management of patients who have clinical signs of CNV and exudative macular degeneration requires consultation with a general ophthalmologist experienced in retinal diseases or retina specialist for FA to determine the eligibility for treatment of the lesion.

#### *e. Loss of Vision*

Vision loss from AMD can develop in one of several different ways. The most frequent cause of significant, acute vision loss is exudative maculopathy, where serous or hemorrhagic detachment of the

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neurosensory retina results from leakage of new vessels in the choroid. Another cause for vision loss is geographic atrophy, characterized by degeneration and loss of RPE. This process damages the overlying photoreceptors and produces a chronic decrease in visual function.

Symptoms of dysmorphopsia, blurred vision (especially at near) or central scotoma that accompany drusen or RPE atrophy should arouse suspicion of CNV. Patients should be instructed to contact their optometrist immediately if they develop new symptoms.

There are few indicators or markers for the progression of nonexudative AMD to the exudative stage. Clinical decision making can be facilitated by considering both the patient symptoms and the ophthalmoscopic appearance of the macula. The following risk factors may be identified when the patient has symptoms of new-onset dysmorphopsia or scotoma:

- Suspicion of CNV
- Neurosensory retinal detachment
- Serous or hemorrhagic RPE detachment
- Vitreous hemorrhage
- Disciform scar in the fellow eye in addition to any of the above.

The following signs of AMD may be identified on the basis of examination findings (in one or both eyes):

- Granular retinal pigmentary changes
- Retinal pigmentary degeneration and atrophy
- Drusen within the macular area accompanied by pigmentary changes.

#### **3. Early Detection and Prevention**

Recent research suggests that the effects of aging on the retina may result from oxidative stress to the RPE brought on by depletion of antioxidant enzymes. It is theorized that antioxidants function as defense mechanisms that convert free radicals into stable compounds before they interact with cell membranes to produce damage.<sup>52</sup> The activity of these antioxidant enzymes decreases with age or from the cumulative

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phototoxic effects of exposure to ultraviolet (UV) radiation and blue light.

Acute exposure to longer wavelength UV radiation and visible light can cause retinal phototoxicity and damage.<sup>34,41</sup> A relationship seems to exist between increased cumulative exposure to longer wavelength UV and visible light and exudative macular degeneration.<sup>53</sup> However, strong epidemiologic evidence for exposure to UVB as a cause of nonexudative AMD is lacking.

The clinician should recommend UV and blue light protection to all patients, but especially children, persons over age 50, and those engaging in vocational or avocational activities requiring increased exposure to UV or visible blue light. These patients should wear spectacles that absorb shorter wavelengths up to visible yellow.<sup>54-56</sup>

The effectiveness of antioxidants in preventing or slowing the effects of AMD is currently being investigated.<sup>53,57</sup> The increased consumption of dark green leafy vegetables (e.g., spinach, kale, collard greens) containing carotenoids (lutein and zeaxanthin) may serve to protect the macula from damaging blue light. Individuals who eat five or more servings of dark green leafy vegetables a week significantly lower their risk for AMD.<sup>58</sup>

The results of various clinical studies are helping to determine the efficacy of nutrient supplementation in the prevention or treatment of AMD.<sup>59-61</sup> There is a growing body of evidence that nutritional supplementation may be effective; however, more data are needed to define the most appropriate nutritional and antioxidant therapies.<sup>62</sup>

Dietary supplementation of antioxidants may be recommended for adults who are nutritionally deficient or noncompliant (e.g., vitamins C [200 mg/day], E [100 IU/day], beta-carotene [10,000 IU/day]; selenium [40 mcg/day]) if there are no contraindications such as anemia or concomitant use of coumadin.<sup>61,63</sup> While moderate zinc supplementation ( 45 mg per day) appears to be safe for most individuals, if Alzheimer's disease or Down's syndrome is suspected or confirmed, zinc supplementation should be avoided.<sup>64</sup> The use of oral zinc sulfate does

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not appear to have an effect on the course of age-related macular degeneration in patients who have an exudative form of AMD.<sup>65</sup> Consultation with the patient's primary care physician may be appropriate before recommendation of nutritional supplements.

## **II. CARE PROCESS**

This Guideline describes the optometric care provided to a patient with age-related macular degeneration. The components of patient care described are not intended to be all inclusive because professional judgment and individual patient symptoms and findings may significantly impact the nature, extent, and course of the services provided. Some components of care may be delegated.

### **A. Diagnosis of Age-Related Macular Degeneration**

The evaluation of patients with retinal changes suggestive of AMD or patients with diagnosed AMD may include, but is not limited to, the following areas.

#### **1. Patient History**

A patient may be unaware of the presence of retinal changes leading to AMD. Risk characteristics for the condition are often determined on the basis of an eye and vision examination. Common ocular symptoms of early AMD include a slow, insidious onset of blurred vision, particularly in the case of atrophic maculopathy. Sudden onset of dysmorphopsia or scotoma indicates a CNVM and attendant retinal complications, most notably serous or hemorrhagic RPE or neurosensory retinal detachment. Vision loss closer to fixation indicates a more central neovascular lesion. Choroidal neovascular membranes can sometimes be located under the center of the FAZ.

#### **2. Ocular Examination**

A number of procedures may be included in the ocular examination for the patient who has or is suspected of having AMD:

- Best corrected visual acuity, including near monocular visual acuity
- Amsler grid testing
- Sensorimotor examination
- Refraction

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- Biomicroscopy
- Tonometry
- Stereoscopic fundus examination with pupillary dilation.

Pupillary dilation with 0.5-1.0% tropicamide, combined with 2.5% phenylephrine or 1% hydroxyamphetamine,\*\* is recommended to achieve maximum stereoscopic visualization of the macula and surrounding retina.<sup>66</sup> Stereoscopic examination of the retina includes fundus biomicroscopy using a hand-held condensing lens (60 D, 78 D, 90 D, Hruby lens, or Superfield NC lens) or universal or other fundus contact lens. Examination of the macula should also involve use of the direct and binocular indirect ophthalmoscopes. All fundus-related procedures should be done through a maximally dilated pupil, unless the use of mydriatics is contraindicated. It is advisable to note and quantify the approximate number, distribution, and appearance of drusen and the extent of RPE abnormalities. Color fundus photography is useful for documenting retinal abnormalities. Red-free retinal photography can help determine the presence and extent of a CNV.

#### **3. Supplemental Testing**

Additional procedures to test for AMD may include, but are not limited to:

- Macular function assessment (e.g., contrast sensitivity, photostress test)
- Color vision
- Central 10-degree computerized automated perimetry
- Fundus photography (including the use of a red-free filter).
- Scanning laser ophthalmoscope

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\* Every effort has been made to ensure that the drug dosage recommendations are appropriate at the time of publication of the Guideline. However, as treatment recommendations change due to continuing research and clinical experience, clinicians should verify drug dosage schedules with information found on product information sheets.

**B. Management of Age-Related Macular Degeneration**

The extent to which an optometrist can provide medical treatment for age-related macular degeneration may vary, depending on the state's scope of practice laws and regulations and the individual optometrist's certification. Treatment of the patient with AMD may require consultation with or referral to the patient's primary care physician, an ophthalmologist, or other health care practitioner for those services outside the optometrist's scope of practice.

A nationwide study was initiated in 1979 to determine the efficacy of laser photocoagulation treatment for specific forms of AMD. This multicenter study had several components:

- The Macular Photocoagulation Study<sup>9,67,68</sup> prospectively evaluated the benefit of laser treatment in preventing severe vision loss in patients with well-defined, extrafoveal CNVM no closer than 200 microns from the center of the FAZ. The criteria for patient selection were established on the basis of patient age; included were persons over age 50 with reasonably good visual acuity and no associated hemorrhage. The MPS reported a noticeable benefit from laser surgery when treating lesions meeting these criteria. Treatment delayed or prevented significant vision loss, especially 1 year after treatment (Table 3).
- The MPS evaluated patients with new and recurrent subfoveal CNV to determine the benefit of treatment for patients who have subfoveal CNV.<sup>68,69</sup>

Although the subjects were patients who had already suffered significant vision loss, the MPS demonstrated that laser photocoagulation had prevented some additional vision loss 2 years after treatment.

**Table 3  
Results of the Macular Photocoagulation Study  
for Extrafoveal Membranes**

VA < 20/200	Treated Eyes	Untreated Eyes
After 1 year	14%	42%
After 18 months	25%	60%
After 3 years	47%	62%

**1. Basis for Treatment**

Management of the patient with nonexudative AMD varies considerably from that of the patient diagnosed with exudative AMD, for whom immediate treatment is critical (See Appendix Figure 1).

**a. Nonexudative AMD**

Patients who have nonexudative retinal changes in AMD may experience no symptoms or may notice slowly progressive changes in visual function (See Appendix Figure 1). A person who has minimal RPE changes and drusen should be evaluated yearly. Patients should be educated and instructed to look for signs of decreased vision, scotoma, and dysmorphism by covering each eye and assessing visual function monocularly. Self-assessment with the home Amsler grid or similar



method may provide additional information regarding the status of the macula. Progression of the maculopathy is characterized by increased pigmentary atrophy that appears clinically as an increased lightening of the affected area of the retina. Choroidal vessels are often visible through the RPE atrophy. The lesion continues to be well-demarcated, and CNV may develop in a small percentage of cases.

All patients who have geographic atrophy should be evaluated every 6 months to rule out long standing PED as a cause of the atrophic changes. The clinician should instruct all patients with geographic atrophy to return for further examination within 24 hours of the onset of new symptoms. Immediate referral to a retina specialist, if available, or a general ophthalmologist experienced in retinal disease for consultation and FA should be made whenever CNV is suspected or there is new onset of visual symptoms.

Patients deemed stable on the basis of reporting no noticeable change in visual function should be examined yearly. The ocular examination protocol includes the evaluation procedures discussed above. The prescription of antioxidant nutrient supplements and UV and blue light protective spectacle lenses should be considered for patients with nonexudative AMD (See Appendix Figure 2).

**b. Exudative AMD**

Patients at risk for severe vision loss from exudative AMD have CNV and symptoms that include new-onset dysmorphopsia and scotoma, sometimes associated with blurred near vision. Leakage from the CNV will result in a neurosensory, RPE, or combined serous and hemorrhagic detachment. Location of the CNV is critical because lesions with borders located beneath the center of the FAZ portend the worst prognosis. Seventy percent of patients with subfoveal CNV have visual acuity worse than 20/200 two years after observation.<sup>16</sup> An immediate consultation with a retina specialist, if available, or a general ophthalmologist experienced in retinal disease should be obtained when clinical signs and/or symptoms of CNV are present.

Fundus FA should be done to determine whether the patient would benefit from laser treatment of extrafoveal or subfoveal CNVs. Timely intervention may reduce the risk for significant vision loss from recurrent CNV. Because of the rapid growth of CNVMs the fluorescein angiogram should be obtained no sooner than 72 hours prior to treatment. The patient should be evaluated 2 weeks post-treatment. A repeat fluorescein angiogram may be needed to determine the efficacy of treatment, to determine if the entire CNV was obliterated, and to rule out recurrence of CNV. When no further leakage is detected, the patient should be evaluated again in 6 weeks, then every 3 months for 1 year, and every 4-6 months thereafter. This surveillance schedule is necessary to assess the risk for recurrence and, on the basis of risk characteristics of soft confluent drusen and retinal pigment abnormalities, to predict the likelihood of CNVM formation in the fellow eye (See Appendix Figure 2).

Signs of CNV recurrence include:

- Decrease in visual acuity
- New scotomas or dysmorphopsia
- New areas of focal hypopigmentation at the edge of an area of laser treatment
- Persistent subretinal or sub-RPE fluid
- New specks of subretinal blood.

Patients who have cardiovascular disease or hypertension, or who are current cigarette smokers, should be monitored more closely due to the higher frequency of CNV recurrence in these populations. Antioxidant nutrient supplements and UV and blue light protection may be recommended. These measures offer additional protection to the RPE while allowing the patient to participate in the care process.

**2. Patient Education**

Patients should be provided information about their diagnosis and the prognosis of their condition. The clinician should inform patients about the symptoms of AMD and encourage them to be compliant by returning for evaluation immediately when they have new symptoms of blurred

vision, dysmorphopsia, or scotoma. Patient education and compliance are critical in identifying symptoms of disease progression. Self-testing with the Amsler grid often fails to detect abnormalities when used in the home environment.<sup>70</sup> Thus careful explanation of home use of the Amsler grid or other forms of monitoring is important in detecting subtle changes and variations in vision.

The optometrist should have a thorough understanding of the indications and benefits of laser surgery and take a realistic approach with regard to the prognosis of each individual patient. Patients eligible for laser surgery should be told that although photocoagulation may not cure the maculopathy, the treatment may prevent further serious vision loss by obliterating the neovascular lesion.

### **3. Prognosis**

The progression of vision loss in nonexudative AMD is variable and must be evaluated on an individual basis. The ophthalmoscopic appearance of the macula cannot be directly correlated with the degree of visual acuity loss. Foveal involvement appears to occur early in the atrophic process, but the average interval from first observation to legal blindness is 9 or 10 years.<sup>71</sup>

The Macular Photocoagulation Study demonstrated the benefits of timely intervention by laser photocoagulation in cases of extrafoveal membranes in exudative AMD and in the treatment of subfoveal membranes. Most patients with AMD will never develop the exudative form of the disease. Only 10-15 percent of persons with drusen will develop signs of CNV. The incidence of involvement of the fellow eye is estimated to be 28-36 percent during the first 2 years, and the annual rate of bilaterality is about 6-12 percent per year for the next 5 years.<sup>6,15</sup> Risk factors for development of exudative maculopathy in the fellow eye include the presence of large, confluent drusen and RPE hyperpigmentation.

### **4. Management of Patients with Severe, Irreversible Vision Loss**

Patients with AMD are at risk for permanent vision loss and loss of central visual function. Although total blindness from AMD is rare, other unrelated ocular abnormalities may impair peripheral vision. Patients with vision loss often require low vision rehabilitation. Consultation with an optometrist who has advanced training or clinical experience in low vision is advisable because patients may benefit from low vision rehabilitation including the use of specialized optical devices and training.<sup>72,73</sup>

Permanent vision loss ranging from 20/25 to 20/800 may be experienced by patients with AMD. This loss of visual function is often accompanied by or exacerbated by other impairments, such as distortion, fixation loss, poor tracking skills, poor fusional capability, difficulty with low contrast targets, or diplopia. These problems may cause significant additional handicaps for the patient with AMD. The patient may experience loss of independence, loss of employment, loss of socialization opportunities, depression and anxiety, and an overall decrease in the quality of life due to AMD.

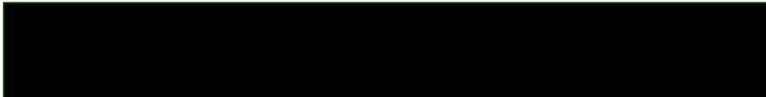
Patients should be evaluated to determine the potential benefits from comprehensive low vision rehabilitation which reduces the debilitating effects of vision loss from AMD. This task-oriented evaluation may include, but is not limited to:

- Expanded patient history and needs assessment
- Evaluation of ocular health
- Low vision assessment of visual acuity (including eccentric viewing)
- Low vision refraction
- Binocular function assessment
- Supplemental testing, including visual fields, contrast sensitivity, and color vision
- Response to optical and electro-optical magnification
- Response to selective absorption filters.
- Response to nonoptical devices (e.g., large print).

Once appropriate optical requirements have been determined, the clinician should educate and train the patient in methods of improving visual function with and without optical devices. The patient should be encouraged to use prescription optical devices for work, home, and social activities.

The goal of low vision rehabilitation is to reduce ocular morbidity and enhance the quality of life. In addition to optical intervention, the evaluation should include the need for nonoptical devices, special lighting, posture aids, contrast enhancement, enlarged print, and nonvisual methods or devices when appropriate. These devices, which significantly enhance the rehabilitative process, are necessary to complement the use of optical devices.

When indicated, the optometrist should recommend blind rehabilitation, occupational, vocational and independent living counseling services and psychosocial consultation. Patients should be informed of other resources including agencies that register and provide services and advocacy to individuals with legal blindness or visual impairment. These agencies can provide information regarding large print and talking books, independent travel aids, and other devices designed to improve quality of life and functional ability within the patient's household. The optometrist should provide the patient written documentation of his or her status relating to legal blindness for state and federal (Internal Revenue Service) tax requirements. Local and national AMD support groups assist many patients in coping with the anxiety and concerns of vision loss. Such groups also provide information regarding resources to help patients function safely and productively in their environment.



## CONCLUSION

Age-related macular degeneration is an important cause of significant vision loss in adults in the United States. In terms of the economic impact, loss of productivity, and quality of life, the disease may be devastating. Until the means to prevent or cure AMD are in place, optometrists need to understand the etiopathogenesis of the disease in order to reduce patient's risk for severe vision loss by early identification and timely referral for laser photocoagulation. Optometrists should educate and inform patients about the natural history of the retinal abnormalities associated with AMD. In certain cases useful vision can be maintained if the patient is informed and educated to seek care promptly. Improved patient understanding of AMD will promote compliance and in some cases may help preserve useful vision.

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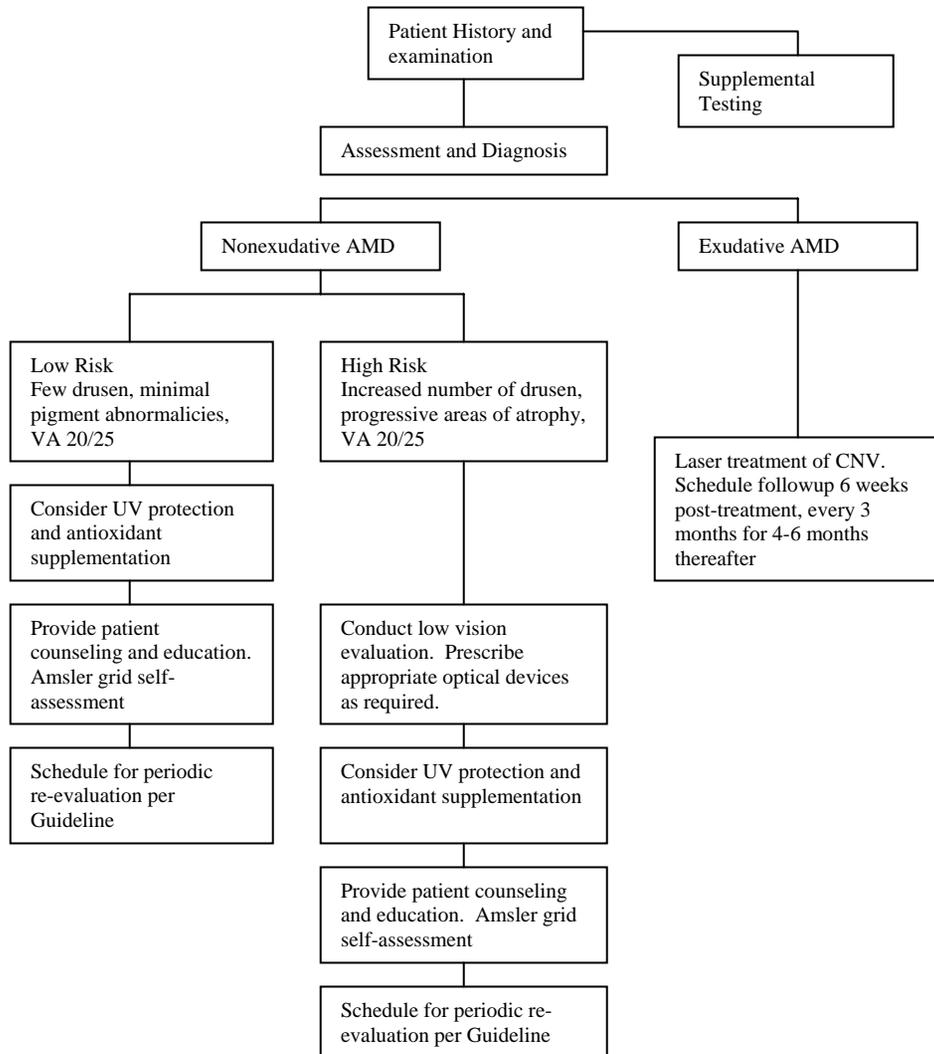
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IV. APPENDIX

**Figure 1**  
**Optometric Management of the Patient with AMD:**  
**A Brief Flowchart**



**Figure 2**  
**Frequency and Composition of Evaluation and Management Visits for Age-Related Macular Degeneration**

<u>Type of Patient</u>	<u>Frequency of Examination</u>	<u>Amsler Grid</u>	<u>Stereo Fundus Biomicroscopy</u>
Patient with two or more risk factors for AMD, over age 55	Annual examination	Yes	Yes
Patient with hard drusen and/or pigmentary degeneration.	6-12 months depending on risk	Yes	Yes
Patient with geographic atrophy, VA 20/30 – 20/70	6-12 months depending on extent of atrophy	Every interim visit	Every interim visit
Patient at high risk with soft confluent drusen and granular pigmentary degeneration	4-6 months	Every interim visit	Every interim visit
Patient with CNV within 2500 microns of center of FAZ	2 weeks after FA laser photocoag-ulation; at 6 weeks, then every 2-3 months after repeat FA	Every interim visit	Every interim visit
Patient with disciform scar in both eyes	6-12 months	Not necessary	Every interim visit

\* Figure 2 extends horizontally through page 41.

**Figure 2 Continued . . .**

<b>Central 10 degree Automated Visual Field (AVF)</b>	<b>Fundus Photography</b>	<b>Management Plan</b>
Yes; baseline, repeat every 2 years	Yes; baseline, repeat every 2 years or as necessary	Patient education; Recommend UVR protection, antioxidant supplementation, home Amsler weekly
Yes; repeat every 2 years	Yes; repeat every 2 years	Patient education; Recommend UVR protection, antioxidant supplementation, home Amsler twice each week
Every 1-2 years	Yes; repeat every year	Patient education; Recommend UVR protection, antioxidant supplementation, home Amsler every other day; Monitor for CNV
Annually	Annually	Patient education; Recommend UVR protection, antioxidant supplementation, home Amsler daily; Low vision consultation and evaluation
Semiannually	Semiannually	Patient education; Recommend UVR protection, antioxidant supplementation, home Amsler daily; Immediate consultation for signs of recurrent CNV; Low vision consultation and evaluation
Annually; consider central 30° AVF, depending on central fixation	Annually	Review; Low vision consultation and evaluation

**Figure 3  
ICD-9-CM Classification of Age-Related Macular Degeneration**

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Drusen (degenerative)	362.57
Exudative senile macular degeneration Kuhnt-Junius degeneration Senile macular degeneration: disciform wet	362.52
Macular degeneration (senile), unspecified	362.50
Nonexudative senile macular degeneration Senile macular degeneration: atrophic dry	362.51
Retinal exudates and deposits	362.82
Retinal neovascularization NOS Neovascularization choroidal subretinal	362.16
Senile reticular degeneration	362.64
Serous retinal pigment epithelial detachment Exudative detachment of retinal pigment epithelium	362.42



**Abbreviations of Commonly Used Terms**

AMD	- Age-related macular degeneration
AVF	- Automated visual field
CNV	- Choroidal neovascularization
CNVM	- Choroidal neovascular membrane
D	- Diopter
FA	- Fluorescein angiography
FAZ	- Foveal avascular zone
MPS	- Macular Photocoagulation Study
PED	- Retinal pigment epithelial detachment
POHS	- Presumed ocular histoplasmosis syndrome
RPE	- Retinal pigment epithelium
UV	- Ultraviolet

**Glossary**

**Amsler grid** A chart with perpendicularly intersecting lines for testing the central visual field.

**Antioxidants** The nutrients beta-carotene (a precursor to vitamin A), vitamin C, and vitamin E that neutralize free radicals implicated in the etiopathogenesis of age-related macular degeneration

**Choroidal neovascularization (CNV)** The proliferation of fragile, newly formed blood vessels originating in the choroidal space and penetrating through to the outer retinal complex into the subretinal and retinal tissue. Serous or hemorrhagic leakage from these vessels results in a neurosensory or retinal pigment epithelial detachment.

**Drusen** The hyaline deposits or colloid bodies of Bruch's lamina of the choroid, very commonly present but not always affecting vision. Drusen are most often seen as a consequence of aging, but are found histologically in younger persons.

**Dysmorphopsia** A distortion of vision due to a disorganization of the photoreceptors, often caused by serous or hemorrhagic elevation of the retina or retinal pigment epithelium. If objects appear larger, the anomaly is referred to as macropsia; if objects appear smaller, it is referred to as micropsia.

**Foveal avascular zone (FAZ)** The rod-free area of the macula within the fovea centralis comprising an area of 350-500 microns and devoid of retinal blood vessels.

**Laser photocoagulation/laserpexy** A process in which a laser burn is used to affix the retina to the underlying choroids in the treatment of retinal breaks, retinal detachment, and other retinal conditions.

**Macula** The central area of the retina, 3-5 mm in diameter, with the foveal depression in the center.

#### 45 Appendix

**Neurosensory retinal detachment** Separation of the neural tissue of the retina, including the photoreceptors, from the retinal pigment epithelium in older individuals, and very often caused by serous or hemorrhagic leakage from newly formed choroidal vessels.

**Retinal pigment epithelium (RPE)** The dark layer of the retina between the lamina of Bruch and the neurosensory retina. It is rich in melanin and provides metabolic support to the photoreceptors and outer retinal complex.

**Scotoma** An area of partial or complete absence of vision surrounded by an area of relatively normal vision. It can be central, paracentral, or peripheral and is referred to as positive or negative when the affected individual is aware (positive) or unaware (negative) of the scotoma.

**Ultraviolet radiation (UVR)** Radiant energy with a wave band shorter than visible light and ranging from 315-400 nm (UVA), 280-315 nm (UVB) and 200-280 nm (UVC). Absorption of UV radiation by ocular tissue has been described as a contributing factor in the etiopathogenesis of AMD and other ocular abnormalities, including cataract and photokeratitis.

**Vitreous hemorrhage** Extension of blood into the hyaloid cavity of the vitreous. It is often associated with choroidal neovascularization in cases of age-related macular degeneration and other conditions in which there is neovascularization.

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