CHRONIC VISUAL LOSS

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Objectives

As a primary care physician, you should be familiar with the major causes of chronic, slowly progressive visual loss in an adult patient-namely:

1. Glaucoma,
2. Cataract,
3. Macular degeneration
4. Diabetic retinopathy and be able to identify the basic characteristics of each.

In addition, you should be able to:

1. Measure intraocular pressure with a tonometer
2. Evaluate the nerve head, classifying it as normal, glaucomatous, or abnormal but non-glaucomatous.
3. Evaluate the clarity of the lens
4. Evaluate the function and appearance of the macula.

To achieve these objectives, you should learn

- To recognize those characteristics of the optic disc useful in determining whether a given disc is normal or abnormal
- To recognize a cataract and to determine its approximate potential effect on the patient's vision
- To determine whether a cataract is the only cause of a patient's visual loss.
- To examine the macula with the ophthalmoscope and recognize the signs and symptoms of maculopathy

Glaucoma

Relevance; Glaucoma is the second most important cause of blindness in the United States and the single most important cause of blindness in African Americans. If glaucoma is detected early and treated medically or surgically, blindness can be prevented. Most patients with early glaucoma are asymptomatic. The great majority of patients lack pain, ocular inflammation, or halos (luminous or colored rings seen around lights). Much peripheral vision can be lost before the patient notices visual impairment.

Glaucoma is usually insidious because symptoms and noticeable visual field defects occur late in the disease. Visual field defects are characterized by arcuate-shaped scotomas and a contraction of the peripheral field, usually sparing the central vision until late in the disease process. Detection in the early asymptomatic stage requires an active effort. The early detection of glaucoma is important because blindness can usually be prevented, if glaucoma is treated adequately and if treatment is started in time.

Because glaucoma involves elevated pressure in the eye, routine measurement of intraocular pressure is a valuable means of screening for glaucoma. Prolonged

elevation of intraocular pressure can lead to optic nerve damage; therefore, examination of the optic nerve is another way to detect glaucoma. Other disorders, such as a brain tumor, can also cause changes in the optic nerve, making the ability to recognize abnormalities of the optic nerve important in and of itself.

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, where it is drained through the trabecular meshwork to Schlemm' s canal, 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 intraocular pressure of 21.5 mm Hg or less.

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, and constitutes an ocular emergency. The abrupt rise in pressure cause symptoms not found in the insidious form of glaucoma, including pain, nausea, and the visualization of colored halos or rainbows around light. 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.

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.

Relationship of IOP and 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. If pressure in the eye is too high, the result may be that blood is prevented from adequately perfusing the optic nerve. If prolonged, this deficiency can damage the nerve.

Damage to the optic nerve results in visual field loss. Such loss is selective but can become severe and even total over time. Detection of glaucomatous visual loss is accomplished by visual field testing. Visual acuity usually does not suffer initially.

Measurement of intraocular pressure and evaluation of optic nerve appearance can detect potential and actual damage so that proper evaluation and treatment can be initiated.

When to Examine: Tonometry and ophthalmoscopy should be part of every comprehensive eye examination. Particular attention should be given to patients who are predisposed, for example, those with diabetes, patients with a family history of glaucoma, and patients over 40, because the incidence of glaucoma increases with age.

How to Examine: Palpation can detect only very hard and very soft eyes; it is very unreliable in the range of the most common glaucomatous pressures. Intraocular pressure is best measured by tonometry, which may be performed in several ways. Indentation, or Schiotz, tonometry uses an instrument that is inexpensive and widely available. The technique is easy to perform takes only a minute or two, and is painless and safe for the patient. The technique of direct ophthalmoscope is particularly useful in assessing the state of the optic disc. An ophthalmologist evaluating a patient with suspected glaucoma would usually perform perimetry to formally evaluate the visual field. The ophthalmologist also may examine the anterior chamber angle structures using a special contact lens on the topically anesthetized cornea, a technique called Gonioscopy.

How to Interpret the Findings: The appearance of the optic disc can be described generally in terms of its color and the size of its physiologic cup. One commonly used description of the optic disc is the ratio of the horizontal diameter of the cup to that of the disc, or the cup:disc ratio. The size of the cup changes little with aging.

The color of the optic nerve can be important in determining atrophy of the nerve due to glaucoma or other causes. Temporal pallor of the optic nerve 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.

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 bend at the edge of the cup. This is also a useful sign in evaluating the size of the cup. Thus, both color and vessel displacement should be evaluated in determining the size of the cup. A cup larger than one half the size of the disc-a cup:disc ratio greater than 0.5 -is suspect. The larger the cup, the greater the possibility of a glaucomatous optic nerve damage.

The optic discs nearly always appear symmetric between the eyes. Discs that exhibit asymmetry of the cup:disc ratios should arouse suspicion. In some cases, there may be edema of the optic disc (called papilledema when caused by elevated intracranial pressure) and the cup may be reduced or obliterated.

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

- Intraocular pressure over 21.5 mm Hg (i.e., a Schiotz reading of 3.5 units or less with a plunger load of 5.5 g)
- Intraocular pressure not elevated, but a difference of 5 mm Hg or more (i.e., 2 Schiotz scale units) between the eyes
- An optic cup diameter greater than one half of the disc diameter (i.e., a cup:disc ratio of 0.6 or greater)

- One cup significantly larger than that of the other eye

- A family history of glaucoma

- Symptoms of acute glaucoma (refer immediately)

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 persons 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. 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 1.5 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.

It is important to be certain that visual loss is explained fully by cataract and not by other causes, such as glaucoma, macular degeneration, or diabetic retinopathy. Cataract may coexist with these conditions, making assessment more difficult.

Basic Information
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 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 radiation damage**.** 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 myopia because of the increased refractive power of the denser nucleus. As the size of the cataract increases, patients become 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.

Patients with posterior subcapsular cataracts may note a relatively rapid decrease in vision, with glare as well as image blur and distortion. This type of cataract is frequently associated with metabolic causes such as diabetes mellitus and corticosteroid use.

With enough time, all cataracts will lead to a generalized impairment of vision. The degree of impairment may vary from day to day. With yellowing of the lens nucleus, objects appear browner or yellower to the patient than they actually are.

When to Examine:
A patient with decreasing vision requires examination to determine the cause of the visual decrease**.** In testing for the presence of cataract, it is also important to attempt to demonstrate that the retina and optic nerve are healthy and that the visual decrease is due to lens changes only.

If the lens is densely cataractous, the ophthalmoscope will not provide a view of the fundus through the opacity. In this situation, the risk of overlooking retinal or other disease conditions exists, as does the risk of performing surgery for cataract without the assurance that vision loss is due primarily to lens changes. Therefore, to detect fundus changes early, ophthalmoscopic examination should be part of every physical examination. Special attention is given to the macula when a patient reports difficulty with near work, blurred vision, or metamorphopsia (i.e., a wavy distortion of central vision).

How to Examine:
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.

Pupillary responses:
Even an advanced cataract would not produce a relative afferent pupillary defect.

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.

Management or Referral:
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 wants to do. It is important not to assign all visual loss to cataract when other, more serious causes of visual loss may be overlooked.

If a cataract interferes with a patient' s daily pattern of living, that patient may benefit from cataract extraction, which can be performed using many different surgical techniques. Patients commonly undergo a laser treatment after cataract surgery to open an opacified posterior capsule, leading to a popular misconception that a cataract can actually be removed with a laser.

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 aged 52 or older. Because certain types of macular degeneration are treated effectively with laser, 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 laser treatable).

Basic Information

Macular Anatomy

The macula is an oval area situated about 2 disc diameters temporal to the optic disc. (The macula is composed of both rods and cones and is the area responsible for detailed, fine central vision. The central macula is avascular and appears darker than the surrounding retina. The fovea is an oval 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 (i.e., foveal reflex).

Age-Related Macular Changes

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

Drusen are hyaline nodules (or colloid bodies) deposited in Bruch' s membrane, which separates the inner choroidal vessels from the retinal pigment epithelium. Drusen may be small and discrete 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, during retinal or choroidal degeneration in disease states, and as a primary dystrophy.

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

About 20% of eyes with age-related macular degeneration develop subretinal neovascularisation. The extension of vessels from the choriocapillaris into the subpigment epithelial space and eventually into the subretinal space means that a defect has developed in Bruch' s membrane.

The subretinal neovascular net may be associated with subretinal hemorrhage, fibrosis, pigment epithelial degeneration, and photoreceptor atrophy. A hemorrhage may result in acute visual loss. 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 neovascularisation and is mandatory before considering laser surgery. Intravenous injection of fluorescein dye and subsequent retinal examination or 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, the retinal pigment epithelium acts as a physical and optical barrier to fluorescein, and thus angiography facilitates identification of pigment epithelial defects. Indocyanine green is another dye used to demonstrate new vessels.

Age-related changes are almost totally 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, or "getting about," 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 decrease. In assessing a patient with decreased 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 vision:

Visual acuity measurement

Amsler grid testing:
Amsler grid testing is a useful method of evaluating the function of the macula. The test is carried out by having the patient look with one eve at a time at a central spot on a page with horizontal and vertical parallel lines making up a square grid pattern. This grid pattern is usually printed in white against a black background. 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 figures in which the eye can distinguish distortions most easily. With the chart held at a normal reading distance of 30 cm from the eye, the Amsler grid measures 10' on each side of fixation. This allows for an evaluation of 5.36 mm in all directions from the center of the macula (i.e., the fovea). 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 may be 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 exudates, and hemorrhage or neovascularisation are all-important signs to check for in an 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:

- A recent onset of decreased visual acuity
- A recent onset of metamorphopsia, or distortion of central vision
- A recent onset of a scotoma, or blind spot
- Any ophthalmoscopic abnormalities in the appearance of the macula, such as drusen, degenerative changes in the retinal pigment epithelium, exudates, or subretinal neovascular membranes

A patient with metamorphopsia may have drusen in the macula only and not be a candidate for laser treatment, but 20% of eyes with age related macular degeneration develop subretinal neovascularisation. Clinical studies have indicated that argon laser photocoagulation of subretinal neovascular membranes that are not too close to the fovea significantly reduces the central visual loss. Recently visudyne therapy with laser has proved effective in stabilizing vision in a well defined sub-foveal sub retinal neovascular membrane.

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 and should be referred to an ophthalmologist capable of supplying these services. More than 11 million Americans have a vision impairment that interferes with routine activities; 1.5 million are classified as severely visually handicapped. The use of visual aids will allow many of these patients to continue to function independently**.** The appropriate and timely intervention by the low-vision specialist is an important part of patients' rehabilitation and should be considered a continuation of their ongoing medical therapy

Points to Remember:

1. Glaucoma should be suspected when ophthalmoscopy reveals either prominent cupping of the optic discs or significant asymmetry of the cup: disc ratio.
2. The primary indication for cataract extraction in most patients is interference with the daily pattern of living rather than reduction of visual acuity to a particular level.
3. Both laser surgery of neovascular membranes and low-vision aids can be helpful to patients with age-related macular degeneration.