

Chapter 2 - Visual Problems

Leonard Hershkowitz, MD

Houston Neurology Associates

Houston, TX

Visual Loss

Perhaps the most challenging and frequent neuro-ophthalmological problem that one encounters is unexplained visual loss. In assessing vision, one needs to evaluate:

- Visual acuity
- Afferent pupillary defect
- Visual fields
- Fundus

Visual Acuity

In testing visual acuity one is actually assessing central or macular vision. This portion of the retina, which consists entirely of cones, is responsible for color perception and the sharpness and clarity of the images that we see. In testing visual acuity the patient can be examined at a distance of twenty feet with the use of a Snellen chart. Each eye is tested individually. Alternatively, a hand held near card may be used at a distance of fourteen inches. When using the near card, always allow the patient to use their reading glasses. The need for reading glasses, termed presbyopia, usually develops in everyone after the age of 40 years. It is due to weakness of the ciliary muscle, which in turn controls the thickness of the lens. If vision is still defective with use of reading glasses, check vision on the Snellen chart as well.

If you have a patient with unexplained visual loss, potential problems include:

- Optical disturbances
- Refractive error
- Disturbances of ocular media (tear film, cornea, lens, vitreous)
- Disturbance of the retina or central neurologic pathways (neuroretinal disturbance)

Tests for Optical Disturbances

Refractive errors of vision are those that can be corrected with proper lenses (glasses). Disturbances of the ocular media can be of several types:

- The tear film can be reduced in conditions causing "dry eyes" such as Sjögren syndrome, or other collagen vascular diseases.
- Corneal infections (keratitis) and corneal edema are sometimes associated with acute closed angle glaucoma.

HOME

AAN.COM

©2013 American Academy of
Neurology - All Rights Reserved

FOR MORE INFORMATION

 memberservices@aan.com

OR

 (800) 870-1960 • (612) 928-6000

- Lens disorder (cataracts).
- Vitreous disorder (hemorrhage, detachment, etc.).

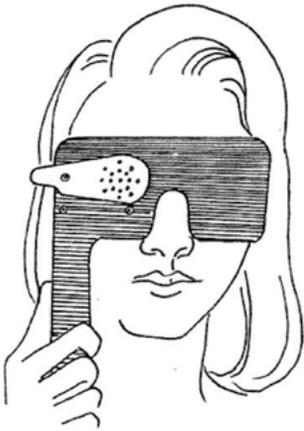


Figure 2-1: Pinhole test

The pinhole test is an effective technique to improve subnormal acuity that is caused by an optical disturbance. One can construct a simple pinhole occluder for office use by taking a piece of cardboard as shown in **Figure 2-1**, and punching a central ~2.5 mm hole, with surrounding holes as shown. If the visual acuity is better with the pinhole than without, then it is much more likely that we are dealing with an optical disturbance. If the acuity is not improved with the pinhole, the examiner should now investigate for a neuroretinal disturbance by searching for a relative afferent pupillary disturbance (RAPD). This abnormality is most commonly seen with disorders of the optic nerve.

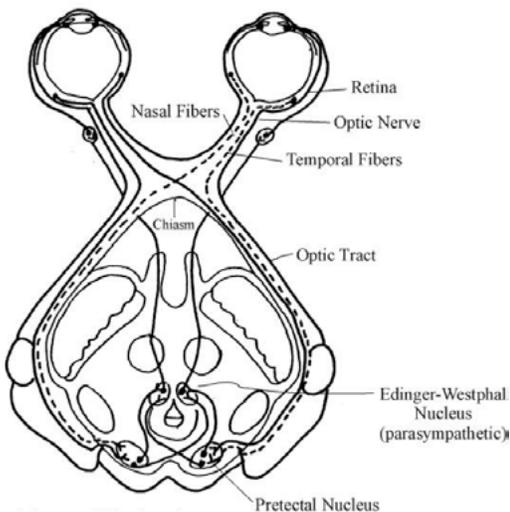


Figure 2-2: Anatomy of light reflex pathway

To understand the concept of the RAPD, we must briefly review the anatomy of the light reflex pathway (see **Figure 2-2**). The pupillary light reflex begins in the retina. Light stimulates photoreceptor cells, which eventually synapse with the ganglion cells. The axons of the ganglion cells that subserve the light reflex undergo hemidecussation in the chiasm (note, in **Figure 2-2**, that only the nasal fibers decussate in the chiasm to the opposite optic tract). Ipsilateral temporal and contralateral nasal fibers continue through the optic tract and then synapse in the pretectal nucleus of the midbrain. Each pretectal nucleus

receives light input from the opposite homonymous visual field. Recall that a homonymous visual field consists of the temporal field in one eye and the nasal field in the opposite eye. Each pretectal nucleus then innervates both Edinger-Westphal nuclei. The result of this arrangement is that unequal light input arising from lesions of the optic nerve or tract does not result in anisocoria (unequal pupils). For example, if one optic nerve is severed both pupils will be of equal size.

To demonstrate the RAPD we perform the swinging-flashlight test (**Figure 2-3**). The patient is seated in a room with very little background illumination. Have the patient fixate on a distant object so as not to invoke a near response (miosis, convergence, and accommodation). Shine a bright light into one eye and observe the direct response (the response to light directly focused on the pupil). Remember, the consensual response (the response of the opposite eye) will be the same as the direct response because of the light reflex pathway. Then quickly shift the light from the first eye to the second one. Under normal circumstances, there should be no change in the pupillary size. **Figure 2-3** is an

HOME

AAN.COM

©2013 American Academy of
Neurology - All Rights Reserved

FOR MORE INFORMATION

 memberservices@aan.com

OR

 (800) 870-1960 • (612) 928-6000

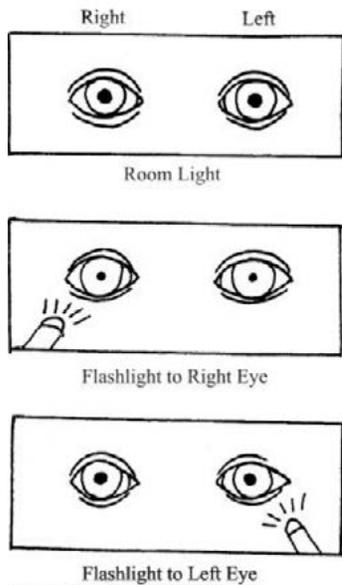


Figure 2-3: Swinging flashlight test

example of findings of the swinging flashlight test in a patient with a defective left optic nerve. Note in panel 2 that both pupils constrict equally. When the flashlight is quickly moved to the defective left eye both pupils dilate equally. And if we were to move the flashlight rapidly to the normal right eye, both pupils would again constrict. Because we are only viewing one pupil at a time, when we move the flashlight to the defective pupil we observe an apparent paradoxical dilatation to light. Actually, of course, both pupils are dilating because of the defective afferent input due to the defective left optic nerve.

In the evaluation of the patient with unexplained monocular visual acuity loss, a RAPD is strongly correlated with ipsilateral optic nerve pathology. Although less likely, it may be observed in retinal nerve fiber layer pathology macular disease, amblyopia, and optic tract pathology. If RAPD is

present, visual fields should be evaluated. If there is no RAPD, and the pinhole test is negative, one needs to consider amblyopia (congenital blindness due to cortical suppression of vision).

Visual Field Testing

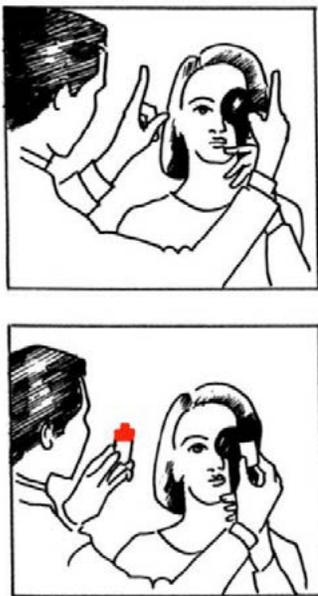


Figure 2-4: Visual field testing

Visual field testing is performed in order to anatomically localize the neuroretinal lesion. In the office setting, confrontational fields are performed. Each eye should be tested individually. The examiner should face the patient and align himself so that he can evaluate the patient's field by comparing it to his own. The patient should fixate on the examiner's eye opposite to his. The index fingers of both hands can be used to check all four quadrants (see **Figure 2-4**). It is imperative to observe whether the field defect respects the vertical meridian (vision is lost or returns when the test object crosses the midline of the patients face. (See **Figure 2-3**; Chapter 1). If it does, the lesion is located at the level of the chiasm or posterior to it. The central field also needs to be examined. This area is especially important in assessing prechiasmatic pathology such as optic nerve or macular dysfunction. These areas are especially sensitive to color desaturation. A red

match tip or red bottle cap can be utilized. The patient with optic nerve disease will frequently have a central scotoma. When the small red object is placed in their central field the color may desaturate and look gray or yellow.

Unexplained Visual Loss

Unexplained loss of visual field with normal visual acuity is usually due to retinal receptor dystrophies and peripheral retinal diseases. Optic neuropathies that

HOME

AAN.COM

©2013 American Academy of
Neurology - All Rights Reserved

FOR MORE INFORMATION

 memberservices@aan.com

OR

 (800) 870-1960 • (612) 928-6000

spare the papillomacular bundle (fibers from the macula to the optic disc that subserve color and central vision) can result in normal acuity. This would include glaucoma, ischemic optic neuropathy, pseudotumor cerebri and rarely, optic neuritis.

Two important facts to note:

Many cases of chiasmatic lesions, and all retrochiasmatic lesions will spare visua acuity.

All retrochiasmatic lesions are homonymous hemianopsias (**Figure 2-3**, Chapter 1).

If one looks at the lesion G in **Figure 2-3**, you will note the hemianopsia has macular or central sparing. This is only seen with occipital lobe lesions because of the large macular representation in the occipital lobe. When observed, this finding is almost always caused by a vascular insult in the occipital cortex.

Fundusoscopic Examination

A thorough examination of the fundus is vital in the evaluation of unexplained visual loss. This is especially true of monocular loss. The disk, macula, retinal vessels, and retina need to be carefully examined. One especially interesting presentation is that of the patient who presents with unexplained visual loss, a RAPD, and a swollen or elevated optic disk. Some of the common etiologies would include optic nerve drusen (congenital hyaline bodies within the optic nerve, sometimes familial in origin), papillitis (inflammation of the optic nerve), ischemic optic neuropathy (arteritic or nonarteritic), and metastatic tumors. Arteritic ischemic optic neuropathy must be diagnosed quickly and accurately. If not treated immediately and aggressively the patient may go blind in a matter of days. This disease is almost exclusively seen in the elderly. There are generally systemic symptoms of myalgia, malaise, anorexia, and headache. Frequently the Westergren sedimentation rate is elevated. In suspicious cases a temporal artery biopsy is indicated; the treatment is high dose steroids.

Pupil Abnormalities

The two most common pupillary problems encountered by the clinician are anisocoria (unequal pupils) and decreased pupillo-constriction to light. The parasympathetic system controls pupillo-constriction. (See **Figure 2-2**, the light reflex pathway). Pupillo-dilatation is controlled by the sympathetic system. The size and shape of the pupil is determined by balance between these two opposing branches of the autonomic nervous system. However, the parasympathetic system is dominant because the iris sphincter is a stronger muscle than the iris dilator.

In evaluating the patient who presents with anisocoria, the clinician should first determine whether the anisocoria is physiological or pathological. Perhaps 20 percent of the adult population will have an inequality of 0.3–0.4 millimeters; only 2 percent will have an inequality of 1 mm or more. If the amount of anisocoria does not change, despite different amounts of background room light and the pupillary reactions to light are equal, physiological anisocoria is the probable diagnosis. The first step in the assessment of pathological anisocoria is to determine whether the inequality is greatest in dim light or in bright light.

If the anisocoria is increased in a darkened room, one may conclude that the

HOME

AAN.COM

©2013 American Academy of
Neurology - All Rights Reserved

FOR MORE INFORMATION

 memberservices@aan.com

OR

 (800) 870-1960 • (612) 928-6000

dilator muscle is deficient. The most common etiology would be a Horner's pupil, due to a sympathetic system deficiency. First, inspect carefully for ptosis. A newly acquired Horner's syndrome in an adult must be thoroughly investigated inasmuch as there is a significant possibility that there is an underlying malignancy. The clinician must assess the structures along the sympathetic pathway including the carotid artery, the apex of the lung, and the base of the brain.

If the anisocoria is greatest in bright light one could conclude that there is dysfunction of the iris sphincter. The differential diagnosis includes disturbances of the parasympathetic system such as Adie's tonic pupil, third nerve palsy, and mydriasis due to pharmacological instillation of drugs that pharmacologically dilate the pupil, such as atropine. Other possibilities include iris damage and acute angle glaucoma causing iris ischemia and subsequent iris sphincter paresis.

Case 1

A 25-year-old female presents with mild headache, and a newly noted large left pupil. Examination demonstrates the inequality of the pupils is increased in bright light, suggesting deficient iris constriction of the left eye. Adie's pupil, a defect in the ciliary ganglion, is a definite possibility. This entity includes:

- relative mydriasis in bright illumination, poor to absent light reaction.
- a slow contraction to prolonged near effort.
- a slow re-dilatation after completion of the near effort (the near stimulus is removed).

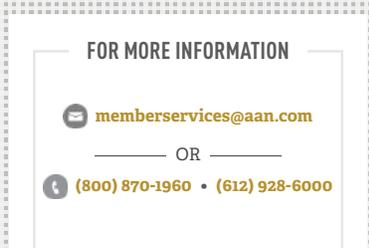
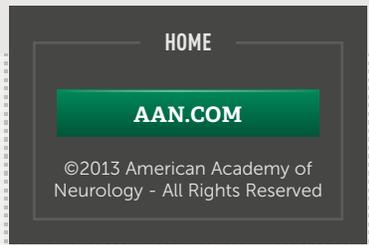
This condition demonstrates light-near dissociation, in which the pupil constricts better to near stimulus, than to light stimulus. It is most frequently seen in women, and when associated with hypo- or areflexia is termed the Holmes-Adie syndrome. Adie's syndrome is generally benign. Further examination fails to demonstrate light-near dissociation. The above patient is now examined for third nerve palsy.

Third Nerve Palsy

One needs to examine ocular motility as well as ptosis. In the absence of those findings, it would be extremely rare for an ambulatory patient to present with a third nerve palsy manifested solely as an isolated dilated pupil. If there is a suspicion that the patient has instilled an atropine-like substance into the eye, one can confirm this by instilling 1 percent pilocarpine solution into the dilated eye. If there is no constriction than one may assume that a mydriatic has previously been in contact with that eye. In a third nerve palsy, the pupil will constrict to the pilocarpine. If the above techniques fail to provide the diagnosis, the patient should be referred to an ophthalmologist in order to inspect the iris with a slit lamp and measure the intraocular pressure. Glaucoma, 2° to acute angle block, is a medical emergency that may present with headache and a fixed pupil.

Ocular Motility

The final portion of this discussion will be devoted to the evaluation of the patient with ocular motility problems, complaining of diplopia.



First, determine whether the diplopia is monocular or binocular. If the diplopia is still present with one eye covered, it is monocular. The common causes of monocular diplopia include cataract, refractive error, corneal abnormalities, retinal surface abnormalities, and psychogenic. If one pinholes the patient and the diplopia clears, one can assume that the etiology was optical. Binocular diplopia is most common; the ocular misalignment is such that if the patient closes either eye the diplopia will resolve. The clinician can get some clues as to the misalignment by asking a few pertinent questions. Are the images separated horizontally, vertically, or obliquely? In what direction of gaze is the separation maximal?

Let's begin our evaluation with a patient that presents with horizontal diplopia. Upon further questioning we learn that the diplopia is worse in gaze to the right. This suggests that there is a defective right lateral rectus muscle or a defective left medial rectus muscle. These two muscles are responsible for horizontal ocular movement to the right. To determine which muscle is responsible, the examiner should check ocular excursions. If, for example, the right lateral rectus fails to fully abduct on gaze right, than the right lateral rectus is deficient. More commonly however, the ocular misalignment is more subtle, and other examination techniques are necessary.

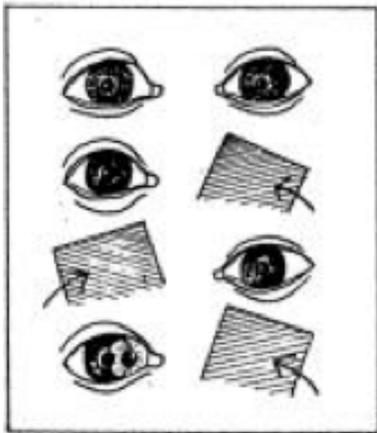


Figure 2-5: Cover-uncover test

One such technique is the alternating cover-uncover test (**Figure 2-5**). The cover-uncover test is based on evoking a fixational eye movement. Remember, if there is ocular misalignment, only one of the eyes fixates on the object of regard while the other eye deviates. Therefore, if the fixating eye is covered, the deviating eye must refixate in order to be aligned with the object of regard. In the cover-uncover test, have the patient fixate on a distant object, then cover one eye. Observe whether the uncovered eye makes a fixational movement, and if so what is the direction of the movement? If no movement is seen, remove the occluder, and place it front of the other eye.

Again observe for any fixational movements of the uncovered eye.

Some common causes of an abduction deficit include:

- Sixth nerve palsy
- Myasthenia gravis
- Extraocular myopathy
- Duane's syndrome
- Spasm of the near reflex

The most common extraocular myopathy is Graves' disease. Duane's syndrome is a congenital condition, felt to be a miswiring of the brainstem. On adduction of the involved eye, the palpebral fissure narrows secondary to retraction of the globe. Interestingly, these patients generally do not complain of diplopia. Spasm of the near reflex is usually secondary to psychological circumstances, and invokes the near triad of convergence, miosis, and accommodation.

HOME

AAN.COM

©2013 American Academy of
Neurology - All Rights Reserved

FOR MORE INFORMATION

 memberservices@aan.com
 OR
 (800) 870-1960 • (612) 928-6000

Convergence mimics the abduction deficit, i.e., the patient converges as you ask them to look left to right, giving the impression that they cannot abduct.

Testing for the specific muscle imbalance in vertical diplopia is much more complex than horizontal diplopia. There are a total of eight muscles, the recti and the obliques, any of which can be responsible for vertical diplopia. In addition, one can have vertical diplopia secondary to skew deviation, a central nervous system abnormality. For those interested in the evaluation of vertical diplopia, please consult the many standard ophthalmological and neuro-ophthalmological texts.

This brief chapter provides the examiner with a basic approach to the evaluation of patients presenting with visual, pupillary, or ocular motility symptoms, as well as some of the more common etiologies. It is imperative that the clinician familiarizes himself with the basic neuroanatomy of these systems.

REFERENCES

Glaser JS. Neuro-ophthalmology. 3rd. Ed. Philadelphia: Lippincott Williams & Wilkins, 1999.

Kline LB, Bajandas FJ. Neuro-ophthalmology review manual. 5th Ed. Thorofare, NJ: Slack, 2001.

Slamovits TL, Burde RM. Neuro-ophthalmology. Textbook of Ophthalmology, Vol. 6. Louis: Mosby-Year Book, 1994.

MULTIPLE CHOICE QUESTIONS

- The pinhole test may improve subnormal acuity caused by:
 - Cataract.
 - Retinal detachment.
 - Optic neuritis.
 - Occipital lobe lesions.
 - None of the above
- A relative afferent pupillary defect is seen most commonly with:
 - Macular degeneration.
 - Vitreous hemorrhage.
 - Amblyopia.
 - Optic neuropathy.
 - None of the above.
- Physiological anisocoria may be seen in what percent of the adult population?
 - 10%
 - 20%
 - 30%
 - 40%
 - None of the above

HOME

AAN.COM

©2013 American Academy of
Neurology - All Rights Reserved

FOR MORE INFORMATION

 memberservices@aan.com

OR

 (800) 870-1960 • (612) 928-6000

4. An Adie's tonic pupil is sometimes associated with:
 - A. Horner syndrome
 - B. Optic neuropathy
 - C. Color vision abnormalities
 - D. Areflexia
 - E. None of the above
5. Common causes of monocular diplopia include:
 - A. Cataract
 - B. Refractive error
 - C. Corneal abnormalities
 - D. Retinal surface abnormalities
 - E. All of the above
6. Which of the following will not cause an abduction deficit?
 - A. Sixth nerve palsy
 - B. Spasm of the near reflex
 - C. Duane's syndrome
 - D. Ischemic optic neuropathy
 - E. All of the above
7. When testing visual fields by direct confrontation, it is critical to note whether the field defect respects the:
 - A. Horizontal meridian
 - B. Vertical meridian
 - C. Neither
 - D. Both
8. When testing for a Horner pupil, there is a greater discrepancy in the anisocoria in:
 - A. A darkened room
 - B. A well-lit room
 - C. The ambient lighting makes no difference whatsoever
9. (True or False) In a patient with optic nerve disease, the swinging flashlight test will demonstrate a subtle anisocoria.
10. (True or False) When using the swinging flashlight test to demonstrate the relative afferent pupillary deficit, the direct response is usually stronger than the consensual response.

HOME

AAN.COM

©2013 American Academy of
Neurology - All Rights Reserved

FOR MORE INFORMATION

 memberservices@aan.com

OR

 (800) 870-1960 • (612) 928-6000

ANSWERS:

1. A
2. D
3. B
4. D
5. E
6. D
7. B
8. A
9. False
10. False



HOME

AAN.COM

©2013 American Academy of
Neurology - All Rights Reserved

FOR MORE INFORMATION

 memberservices@aan.com

OR

 (800) 870-1960 • (612) 928-6000