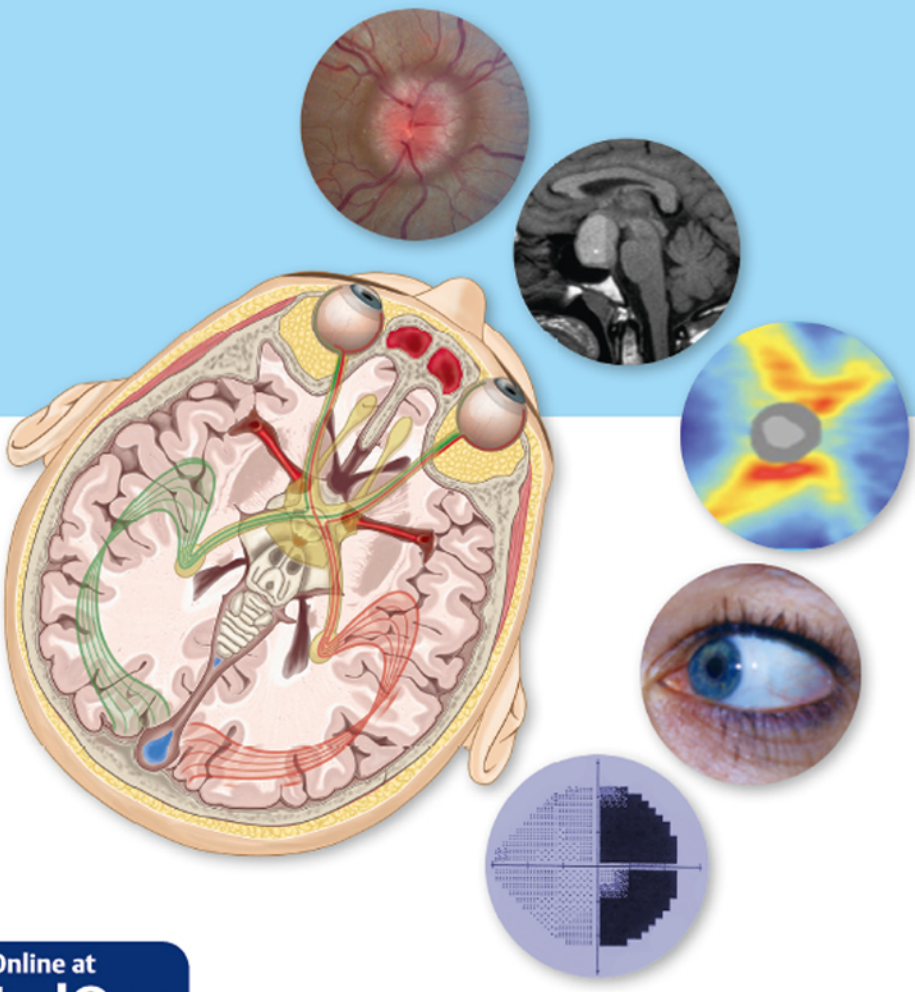
Neuro-Ophthalmology Illustrated

Third Edition

Valérie Biousse
Nancy J. Newman



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In your bag:
– Near card
– Reading glasses (+2.00 or +3.00)
– Pinhole
– Red object (such as a pen, or top of dilating drops)
– Striped ribbon or paper to test optokinetic nystagmus
– Amsler grid
– Short lasting dilating drops
– Direct ophthalmoscope with spare batteries

Visual acuity	Distance		Near	
	Right eye	Left eye	Right eye	Left eye
Without correction				
With correction or pinhole				

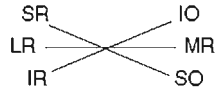
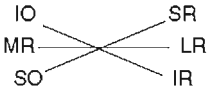
Color vision	Right eye	Left eye

Amsler grid	Right eye	Left eye

External examination	Right eye	Left eye
Orbits		
Lids		

Pupils	Right eye	Left eye
Size in dark (mm)		
Size in light (mm)		
Response to light		
RAPD		
Reaction at near		

Ocular examination (Slit lamp or penlight)	Right eye	Left eye

Ocular motility	Right eye	Left eye
Eye movements		

Fundus	Right eye	Left eye

Visual fields	Left eye	Right eye
Confrontation		
Formal: GVF HVF		

Other cranial nerves	Right	Left

Blood pressure		
Neurological examination		
Other		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We dedicate this book to our students

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Preface to the 3rd Edition

Third time's the charm! Welcome to the 3rd edition of Neuro-Ophthalmology Illustrated. From the moment in 2015 when we passed on the 2nd edition of our book to the copy editor, we have been thinking about how we can make the book even better. We asked this of our trainees, our colleagues, and those who have used this book in the global real-world setting. They answered, and we listened. In the 3rd edition, not only have the text and tables been fully updated, but so too have the illustrations, adding some, updating others, improving their overall quality and layout. But most importantly, the 3rd edition has been enhanced by the incorporation within the text of 69 video clips of examination techniques, normal and abnormal eye movements, pupil findings, and other moving images, all easily accessible within the pertinent sections. We hope you enjoy!

Preface to the 2nd Edition

Over the 6 years since the publication of the first edition, Neuro-Ophthalmology Illustrated has sold by the thousands. Reviews have been spectacular and we are overwhelmed by the positive response worldwide, from both trainees and mentors. It has been lauded as “the best soft-cover training manual in neuro-ophthalmology”, “a tour de force”, and even “a great stocking stuffer for every neurology and ophthalmology resident”. We are very proud and humbled by its success.

We hope now to have made it better. This second edition has been fully updated. Although the overall structure of the book remains the same, new sections have been added and space used more efficiently. Illustrations have been enlarged and clarified, the inevitable errors identified by our eagle-eyed trainees have been corrected, and feedback from our readers has been incorporated. We hope you enjoy!

Preface to the 1st Edition

Neuro-ophthalmology is the "overlap" specialty between ophthalmology and neurology. It covers all the disorders that may affect those parts of the central nervous system devoted to vision: the afferent visual system (the pathways for visual input and processing), which encompasses more than one third of the supratentorial brain mass, and the efferent system (the pathways for ocular motor control and pupillary function), which crisscrosses throughout the brainstem and cerebellum. Indeed, it is hard to imagine a neurologic disorder that could not have neuro-ophthalmic manifestations. Hence, neuro-ophthalmology has also been referred to as "applied neuroanatomy."

Neuro-ophthalmology is also that part of ophthalmology that most scares ophthalmologists and that most confuses neurologists. Ophthalmologists, skilled in the direct inspection of the eye, are accustomed to seeing the pathology, rather than inferring it. Brain tumors, ischemic events, and inflammation can present with the same patient complaints such as refractive error, dry eye, or cataract, but the implications for management and prognosis are dramatically different, and correct diagnosis is essential. Neurologists, although perhaps more comfortable with the diagnosis and management of life-threatening neurologic processes, are often challenged by the techniques of the examination of the visual system-skills frequently neglected in their training and relegated to "eye doctors." Diagnosing brain disease using the eye examination is not easy if the components of the eye examination include skills and techniques unfamiliar to the neurologist.

Perhaps most importantly, neuro-ophthalmology is deeply rooted in a certain way of thinking. It begins first and foremost with neuroanatomy, moves next to the mechanisms of disease, then generates a differential diagnosis of which specific disorders to consider, and finally addresses the appropriate management for diagnosis and treatment: (1) Where? (2) How? (3) What? (4) Now what? For example, in a patient presenting with visual loss, the first step is to localize the lesion along the pathways of vision (Where?). Let's say that the localization is the optic nerve. The second step, then, is to review all those categories of disease that can affect the optic nerve (inflammatory, vascular, compressive, toxic, etc.) and decide which one best fits the clinical profile of the case (How?). Once the likely mechanisms have been identified, specific disorders can be considered (What?). Ultimately, the appropriate diagnostic tests and further management will depend on the preceding process of logical thinking (Now what?).

Welcome to Neuro-Ophthalmology Illustrated. It is hard to imagine a more "visual" subspecialty than neuro-ophthalmology. All the layers of the eye are available for direct inspection using the techniques and tools of the ophthalmologist, and all the features of the brain are exposed by today's exquisite advances in neuroimaging. Over the course of 20 years of teaching medical students, residents, fellows, and practitioners, we have personally acquired over 20,000 unique clinical images. We have used more than 1000 of these images in this book to illustrate the phenomenal richness of clinical neuro-ophthalmology. More than an atlas, Neuro-Ophthalmology Illustrated aims to provide the essential information on basic clinical neuro-ophthalmology and to simplify the perceived complexity of neuro-ophthalmology, without sacrificing comprehensiveness.

This book began with the idea of a small, practical, and portable manual for medical students and residents, especially residents preparing for board examinations in ophthalmology, neurology, and neurosurgery, as well as those pursuing careers in neuroradiology, otolaryngology, and even primary care. We have attempted to compensate for the diversity of our audience by providing the necessary basic information on the two primary specialties that overlap to become neuro-ophthalmology. For the ophthalmology-naive student of neurology, we provide a basic introduction into the anatomy, physiology, and examination of the eye. For the ophthalmologist, we reciprocate with practical examples of brain anatomy and circuitry, as well as the fundamentals of neuroimaging. The book is structured to include sections on the essential components of the neuro-ophthalmic examination and evaluation; disorders of the visual afferent system, pupil, ocular motor efferent systems, orbit, and lid; evaluation of the nonorganic patient; and common or classic neurologic and systemic disorders with important

neuro-ophthalmic manifestations. A more comprehensive index appears at the end of the book. The emphasis is on how to think about these disorders, from symptoms and signs, to localization, differential diagnosis, and management.

Neuro-ophthalmologists are teachers. We teach ophthalmologists about the brain and neurologists and neurosurgeons about the eye. We facilitate communication between specialties and among physicians. Although we believe our book is still the practical manual it was meant to be, we hope that the richness of the illustrations will serve as a reference for practitioners in all the related fields and for our fellow teachers of neuro-ophthalmology We hope that Neuro-Ophthalmology Illustrated facilitates your learning and your teaching.

Acknowledgments

We wish to thank those who have helped us with this third edition. Specifically, we are grateful to William Lamsback, Thieme's Executive Editor for Ophthalmology Publications, and to our Managing Editor, Liz Palumbo, who signed on with us for yet another go. But most importantly, we would like to acknowledge the dedicated hard work and superb video editing of our Video Editor, Jonathan Micieli, MD, who helped transform this 3rd edition into a book of its times.

1 The Neuro-Ophthalmic Examination

Abstract
The neuro-ophthalmologic examination is an integral part of the neurologic and ophthalmologic examinations. Its goal is to detect and localize lesions that involve the visual afferent and efferent systems. Its fundamental components include tests of visual acuity, color vision, eyelid position, orbital appearance, pupillary size and reactivity, ocular integrity, ocular motility, visual fields, and ocular funduscopic appearance.

Keywords: neuro-ophthalmologic examination, visual acuity, color vision, ocular motility, pupils, visual fields, ocular fundus

1.1 Introduction to the Neuro-ophthalmic Examination

A detailed neuro-ophthalmic examination is part of routine neurologic and ophthalmic examinations. It is a powerful means to detect and localize lesions that involve the visual system. Documentation of the extent of damage within the visual system is also an invaluable method to assess the effect of various therapies and often guides the management of numerous neurologic and neurosurgical disorders.

The extent of the neuro-ophthalmic examination varies depending on the patient's complaints, but parts of it should always be performed in detail in selected neurologic disorders, and some parts of the neuro-ophthalmic examination should be systematically performed in most neurologic and systemic diseases. For example, in a patient with an occipital infarction, evaluations of visual acuity, color vision, and formal visual fields are the most important. A patient with known multiple sclerosis needs a thorough examination, because all functions involved in the visual system may be affected. In a patient complaining of diplopia or with anisocoria, formal visual field testing is usually not necessary, whereas all patients with raised intracranial pressure and papilledema should have formal visual field testing, even when they have no visual symptoms.

Most examination techniques detailed here are best performed with appropriate tools in a neuro-ophthalmologist's or ophthalmologist's office. However, a basic neuro-ophthalmic examination (including evaluation of the vision, pupillary function, ocular motility, and funduscopy) can be performed at the bedside, in the emergency room, or in a neurologist's office with only very few tools (see Box 1.1 for a list of required tools).

Box 1.1 Tools needed for a neuro-ophthalmic examination at the bedside

- Near card to check visual acuity.
- A pair of reading glasses (+2.00 or +3.00).
- A pinhole (made from cardboard or plastic with a few small pinholes).
- A red object, such as a pen or the top from a bottle of dilating drops (used to check for color saturation and for visual fields).
- A striped ribbon or paper to test optokinetic nystagmus.
- An Amsler grid.
- Short-lasting dilating drops.
- A direct ophthalmoscope with spare batteries (used to check the pupils, to perform a penlight examination of the eyes, and to examine the fundus).

The examination usually follows a specific order, as given in the following discussion (e.g., you have to examine the visual acuity before flashing light into the eyes; the pupils need to be examined before drops are placed in the eyes; funduscopic examination is the last part of the examination).

1.2 Visual Acuity

In examining the patient's visual acuity, each eye is tested separately (▶ Fig. 1.1). Visual acuity is measured with the patient's corrective lenses or a pinhole (▶ Fig. 1.2). There are two types of visual acuity tests: distance and near.

Distance visual acuity is tested as follows (▶ Fig. 1.1, ▶ Fig. 1.2):

1. Place a Snellen chart at 20 feet.
2. Record the smallest letters read by the patient with each eye (e.g., "20/20 right eye with correction; 20/25 left eye with pinhole").
3. If the patient cannot read the largest letter, visual acuity is less than 20/800 and is recorded as "count fingers," "hand motion," "light perception," or "no light perception."

The pinhole is an opaque panel perforated with one or more holes 1.0 to 1.5 mm in diameter. The holes restrict incoming light rays to a narrow path that bypasses refractive irregularities and presents a single, focused image to the fovea of the retina. Refractive errors and visual loss from cataracts improve with a pinhole. If visual acuity cannot be improved with a pinhole, then other media opacities, optic nerve disease, maculopathy,

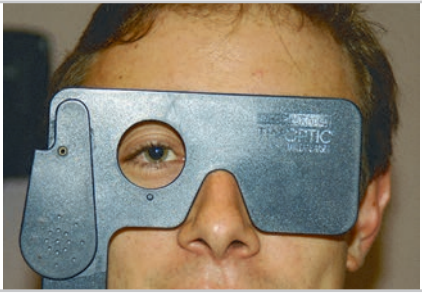

Fig. 1.1 Cover one eye to measure visual acuity. The occluder can be used over the patient's glasses.

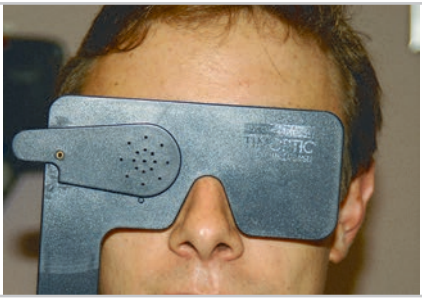

Fig. 1.2 The patient is asked to try to read smaller letters through the pinholes.

Fig. 1.3 Near vision is tested using a near card held at 14 inches from the patient. It must be tested with the patient's correction for near vision. Most patients older than age 50 will need a +2.00 or +3.00 lens to read.

or amblyopia is likely the cause of visual loss. The test may be unreliable with young children, elderly, and cognitively impaired individuals.

Near visual acuity is tested as follows (▶Fig. 1.3, ▶Fig. 1.4):

1. Hold a near card at 14 inches from the patient. The patient should be tested with reading glasses (or a +2.00 or +3.00 lens) if he or she is older than age 50 (because of presbyopia).
2. Record the smallest letters or numbers read by the patient with each eye (e.g., "J1 + right eye with correction; J1 left eye with correction").

1.3 Color Vision and Color Saturation

Color vision can be assessed by several methods, and each eye is tested separately. The purpose of color vision testing is to detect acquired unilateral or bilateral color loss, which occurs most commonly with maculopathies, optic neuropathies, chiasmal disorders, and, more rarely, bilateral occipital lesions.

The Ishihara pseudoisochromatic and the Hardy–Rand–Rittler color plates are routinely used (▶Fig. 1.5). The number of plates correctly identified with each eye is recorded (e.g., "14/14 Ishihara color plates, right eye; control only left eye"). The control plate can be read by patients with a visual acuity of at least 20/400. Some patients with dementia and simultagnosia may have difficulty using these plates.

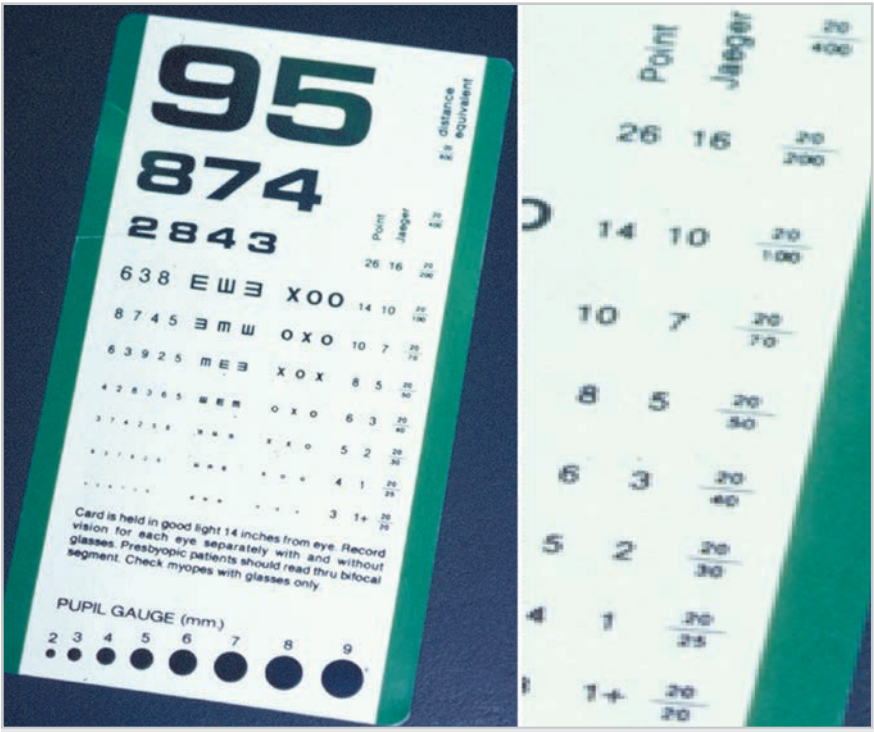

Fig. 1.4 The near card uses numbers or letters. It is usually measured in Jaeger numbers (J1 + corresponds to 20/20, J16 to 20/200).

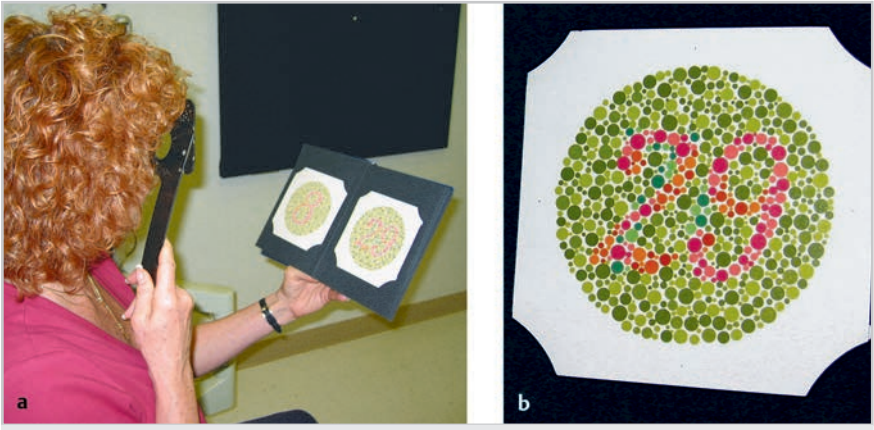

Fig. 1.5 (a,b) Color testing with the Ishihara color plates.

When color plates are unavailable, the difference in color perception between the two eyes may be identified using a red object (e.g., the top from a bottle of dilating drops; ▶Fig. 1.6). Even with normal color plate testing, the patient may recognize a

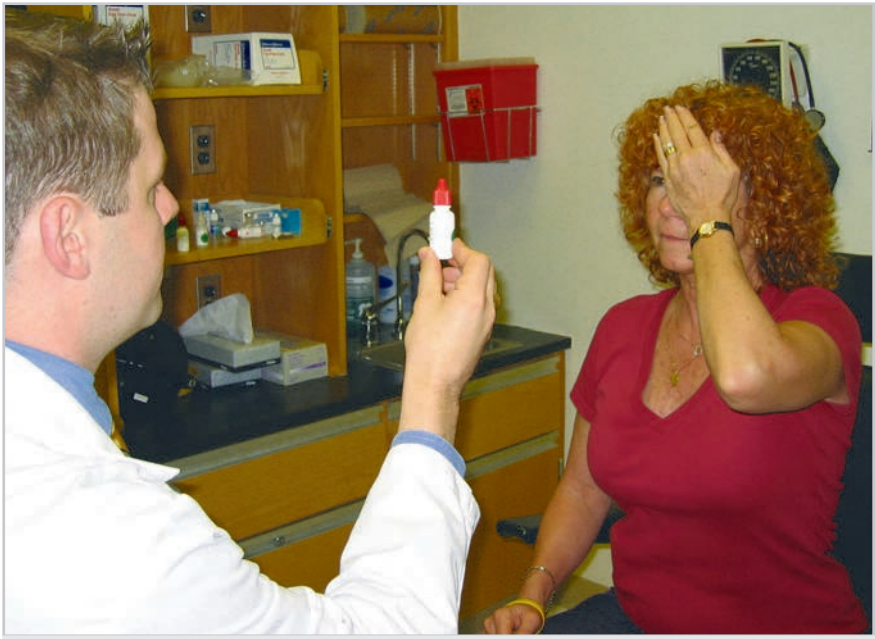

Fig. 1.6 Comparison of color saturation between the two eyes using a red object.

color difference in a red bottle top alternately presented to each eye. The patient should be asked to quantify the red desaturation (percentage of normal).

1.4 Contrast Sensitivity

Contrast sensitivity is another measure of visual function and is often abnormal in patients with optic neuropathies. Patients with maculopathies and cataracts also often have decreased contrast sensitivity. It is not tested in all patients but is useful in patients with visual complaints and an otherwise normal examination. It is also used as a measure of visual function in numerous clinical trials, especially multiple sclerosis trials. The test uses a chart with letters or stripes represented in various shades of gray.

1.5 Photostress Recovery Test

Photostress recovery is used to differentiate between macular disease and optic neuropathy. The principle underlying this test is that recovery of retinal sensitivity following exposure to a bright light is based on regeneration of visual pigments that were bleached during exposure to light. A delay in this process occurs in diseases affecting the photoreceptors and is independent of the neural pathways.

Each eye is tested separately:

1. Measure the best corrected visual acuity in each eye.
2. Have the patient look directly into a bright light held a few centimeters from the eye for 10 seconds.
3. Record the time taken for the visual acuity to return to within one line of the best corrected visual acuity.

Most normal patients will have a recovery time of less than 30 seconds, which is symmetric between the two eyes. Macular diseases (but not optic neuropathies) often cause a prolongation in the photostress recovery time. This is particularly useful for unilateral or subtle macular diseases.

1.6 Amsler Grid

The Amsler grid is very useful in detecting macular abnormalities as a cause of visual loss (▶ Fig. 1.7a).

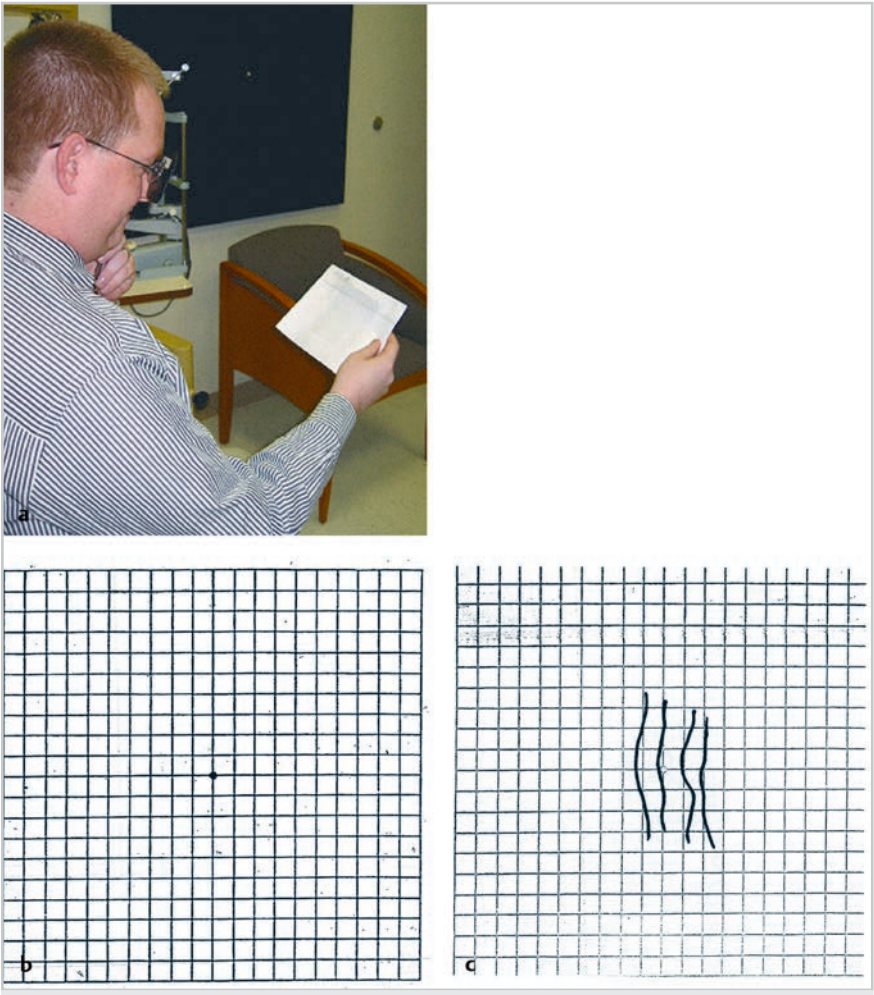

Fig. 1.7 **(a)** Amsler grid testing. **(b)** Normal Amsler grid. **(c)** Amsler grid showing central distortion of the lines.

Each eye is tested separately, and the patient is asked to fixate on a central point in a square grid of lines and to draw any area in which the lines disappear or are broken, warped, double, or curved (▶Fig. 1.7b).

Patients with maculopathy often see the straight lines as curved (metamorphopsia; ▶Fig. 1.7c).

1.7 Stereo Vision

Stereo vision is tested on a specific book (Titmus test) with both eyes open and polarized glasses placed on the patient's reading corrective lenses. This book shows animals and circles that are seen in stereo with the polarized glasses (▶Fig. 1.8).

Stereopsis requires binocular vision. Therefore, the presence of stereopsis indicates at least some vision in each eye. This test is very helpful when nonorganic visual loss is suspected.

Stereopsis can be quantified and grossly correlated with visual acuity (▶Table 1.1, ▶Table 1.2).

Fig. 1.8 Stereo vision testing with the Titmus test.

Table 1.1 Relationship of visual acuity to stereopsis (Levy and Glick scale)

Visual acuity in each eye	Stereopsis (arc seconds)[a]
20/20	40
20/25	43
20/30	52
20/40	61
20/50	89
20/70	94
20/100	124
20/200	160

[a]The Titmus test gives results in seconds of arc.

Table 1.2 Estimate of relation of visual acuity to stereopsis

Titmus Score (circles)	Worse eye visual acuity, 95% prediction interval (Snellen ratio)	Worse eye visual acuity, 99% prediction interval (Snellen ratio)
9	20/40	20/79
8	20/45	20/95
7	20/50	20/117
6	20/62	20/181
5	20/76	20/266
4	20/106	20/568
3	20/176	20/1833
2	20/687	20/39009
1	20/5263	20/7557835

Note: When seeing a patient who claims 20/200 visual acuity in one eye and correctly identifies 9 circles, you can predict the worse visual acuity to be at least about 20/40 with 95% confidence and at least 20/80 with 99% confidence. If the patient identifies 6 circles correctly, the worse visual acuity is at least about 20/60 with 95% confidence and 20/180 with 99% confidence.

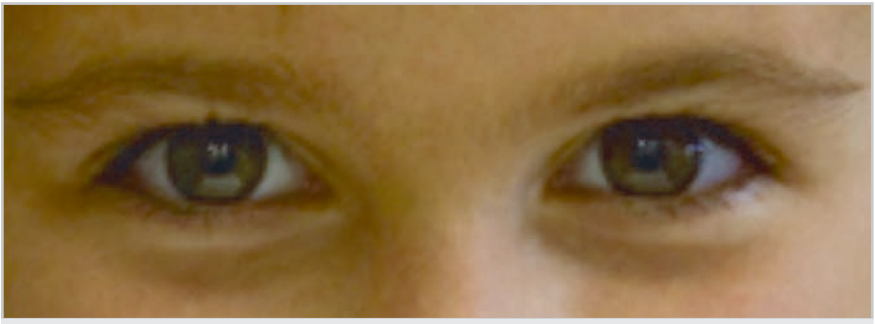

Fig. 1.9 Normal eyelids.

1.8 Eyelid Examination

An eyelid examination includes evaluation of the following:

- Position of the eyelids.
  - Ptosis (droopy eyelid).
  - Retraction.
- Lid function.
- Swelling.
- Mass.

In normal individuals, the upper lid covers the superior 1 to 2 mm of the iris, while the lower lid just reaches the inferior aspect of the iris (▶Fig. 1.9). Examination of the eyelids (▶Fig. 1.10, ▶Fig. 1.11) includes measurements of the following:

- Palpebral fissure: distance between the upper and lower eyelid in vertical alignment with the center of the pupil (normal: 9–12 mm).
- Margin reflex distance (normal: 4–5 mm).

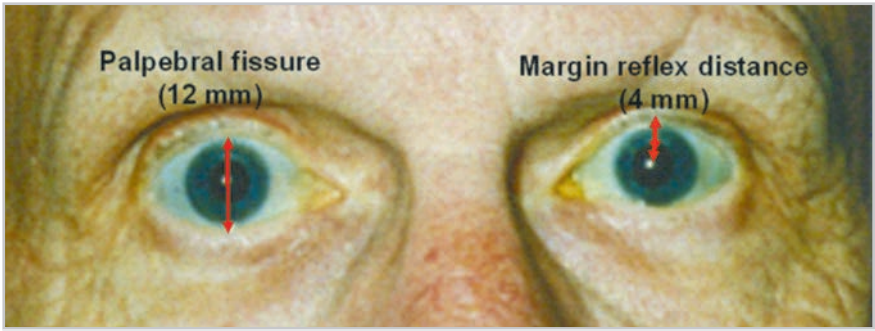

Fig. 1.10 Measurement of palpebral fissure and margin reflex distance.

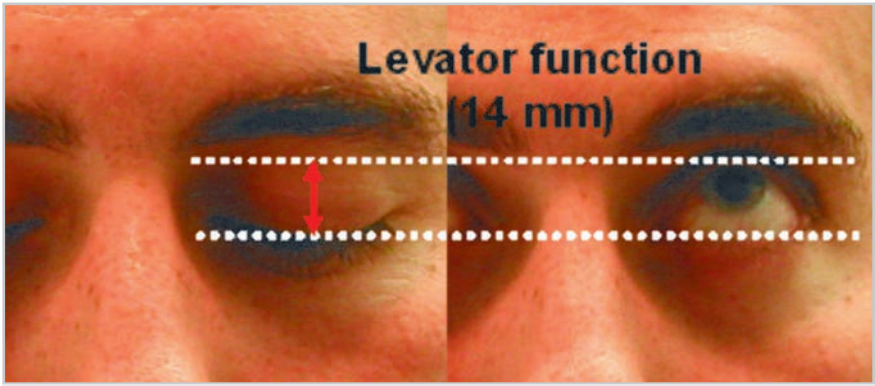

Fig. 1.11 Measurement of levator function.

  - Marginal reflex distance-1: distance between the center of the pupillary light reflex and the upper eyelid margin with the eye in primary gaze.
  - Marginal reflex distance-2: distance between the center of the pupillary light reflex and the lower eyelid margin with the eye in primary gaze.
- Levator function: distance the eyelid travels from downgaze to upgaze, while the frontalis muscle is held inactive at the brow. A measurement of greater than 10 mm is considered excellent, whereas 0 to 5 mm is considered poor.

For more information on disorders of the eyelid, see Chapter 17.

1.9 Orbital Examination

An orbital examination includes the following:

- Inspection of the patient's external appearance.
  - Orbital deformations.
    - Hypo- or hypertropia of the globes.
  - Abnormal position of the eyes within the orbits.
    - Proptosis (eye bulging out of the orbit).
    - Enophthalmos (eye sinking into the orbit).

- Periorbital soft tissues.
  - Swelling.
  - Redness.
  - Hematoma.
  - Mass.
- Palpation of the orbital rims.
- Resistance to retropulsion of the eyes.
- Auscultation of the orbital contents (for a bruit).

Proptosis can be measured with the Hertel exophthalmometer (▶Fig. 1.12) and on neuroimaging (▶Fig. 1.13). Deformations (▶Fig. 1.14) and disease (▶Fig. 1.15) cause various orbital syndromes. For more information on orbital syndromes, see Chapter 14.

1.10 Pupillary Testing

Pupils should be tested in the dark with a bright light and with the patient fixating at a distance (▶Fig. 1.16). Pupil examination includes the following (Table 1.3):

- Size.
- Presence of anisocoria (difference of size between the two pupils).
- Response to light.
- Presence of a relative afferent pupillary defect (RAPD).
- Dilation in the dark.
- Constriction at near.

For a full discussion on the pupil, see Chapter 12 and Video 12.1.

Fig. 1.12 Hertel exophthalmometer.