

Disorders of the Eye

Chapter 34 | Part 2: Cardinal Manifestations and Presentation of Diseases

KEY CLINICAL POINTS

- 1 Pinhole testing improves acuity in refractive errors but not in optic nerve disease.
- 2 PERRLA is redundant if light response is good; check near response if light is poor (e.g., Argyll Robertson, Parinaud's, or Adie's).
- 3 Anisocoria increasing in dim light indicates sympathetic paresis (Horner's syndrome).
- 4 Roth's spots are seen in subacute bacterial endocarditis, leukemia, and diabetes.
- 5 Cherry-red fovea with a milky retina is characteristic of central retinal artery occlusion.
- 6 Amaurosis fugax is a transient ischemic attack of the retina requiring urgent stroke evaluation.
- 7 Herpes zoster ophthalmicus requires antiviral therapy and cycloplegics to prevent scarring.
- 8 Acute angle-closure glaucoma is treated with acetazolamide, beta blockers, and pilocarpine.
- 9 Legal blindness is defined as best corrected acuity of 6/60 (20/200) or less in the better eye.
- 10 Temporal pallor of the optic disc is a more reliable sign of optic atrophy than pallor of the nasal rim.

DEFINITION & CLASSIFICATION

- **Legal Blindness:**

Definition (Harrison's 22e): best corrected acuity of 6/60 (20/200) or less in the better eye or a binocular visual field subtending 20° or less.

- **Optic Atrophy:**

Definition (Harrison's 22e): not a specific diagnosis but refers to the combination of optic disc pallor, arteriolar narrowing, and nerve fiber layer destruction produced by a host of eye diseases, especially optic neuropathies.

ETIOLOGY & PATHOPHYSIOLOGY

Anatomy of the Visual System

- **Retina:**
 - **Function:** Transducer converting light energy into neuronal signals.
 - **Photoreceptors:**
 - Rods: 100 million; function in dim (scotopic) illumination.
 - Cones: 5 million; function in daylight (photopic); specialized for color and high spatial resolution.
 - **Macula:** Contains the majority of cones; serves the central 10° of vision.
 - **Fovea:** Located in the center of the macula; packed exclusively with cones; provides best visual acuity.
- **Neural Pathways:**

- **Ganglion Cells:** 1 million per retina; translate images into action potentials.
- **Optic Nerve:** Contains 1 million fibers; carries signals to the brain.
- **Primary Pathway:** Retinogeniculocortical (Retina → Lateral Geniculate Nucleus → Primary Visual Cortex).
- **Specialized Ganglion Cells:**
 - Melanopsin-containing cells: Govern pupillary constriction and circadian rhythms via pretectal olivary nuclei and the suprachiasmatic nucleus.
- **Motor System (Efferent):**
 - Purpose: Foveation (moving targets onto the fovea).
 - Muscles: 6 extraocular muscles per eye.
 - Innervation: Cranial nerves III, IV, and VI; coordinated by pontine and midbrain centers.

Refractive States

- **Myopia:** Globe too long → focus in front of retina → distant objects blurry.
- Correction: Diverging lens.
- **Hyperopia:** Globe too short → requires converging lens.
- **Astigmatism:** Non-spherical cornea → requires cylindrical corrective lens.
- **Presbyopia:** Age-related loss of accommodation due to lens changes; requires reading glasses or bifocals.
- **Tonic Pupil:**
 - Cause: Seen in multiple system atrophy, segmental hypohidrosis, diabetes, and amyloidosis.
 - Diagnosis: 0.125% pilocarpine → produces constriction in tonic pupils but no response in normal pupils.
- **Gardener's Pupil:** Mydriasis induced by tropane alkaloids (e.g., nightshade, jimsonweed).

CLINICAL FEATURES

Pupillary Examination

- **PERRLA:**
 - Note: 'Near' response is only necessary if light response is poor.
 - Light-near dissociation: Seen in neurosyphilis (Argyll Robertson), Parinaud's syndrome, or Adie's tonic pupil.
- **Anisocoria:**
 - Stable size → essential/physiologic.
 - Increases in dim light → sympathetic paresis (Horner's syndrome: miosis, ptosis, anhidrosis).
 - Increases in bright light → parasympathetic palsy (e.g., CN III palsy).
- **Marcus Gunn Pupil:**
 - Mechanism: Relative afferent pupillary defect (RAPD) during swinging flashlight test.
 - Significance: Useful for retrobulbar optic neuritis and other optic nerve diseases.

Eye Movements & Alignment

- **Saccades:** Rapid, accurate jumps between targets.

- **Assessment:** Patient looks back and forth between two stationary points.
- **Alignment:**
- **Test:** Cover test (detects heterotropia as small as 1°).
- **Measurement:** Prism power used to determine magnitude of deviation.
- **Stereacuity:**
- **Normal:** 40 s of arc.
- **Screening:** Random dot stereograms.

Color Vision

- **Cones:** Three types (Red, Green, Blue) with peaks at 560nm, 530nm, and 430nm.
- **Genetics:** Red/Green on X chromosome; Blue on Chromosome 7.
- **Congenital Color Blindness:** Affects 8% of males (anomalous trichromats or dichromats).
- **Acquired Defects:**
- **Cause:** Macular disease, optic nerve damage, or bilateral strokes (cerebral achromatopsia).
- **Clinical Signs:** Color desaturation, prosopagnosia (difficulty recognizing faces), or color anomia.

DIAGNOSTIC APPROACH

Visual Acuity Testing

1. **Initial Assessment:** Determine if refractive error is present.
 - **Pinhole Test:**
 - **Improvement** → Refractive error → Proceed to refraction.
 - **No improvement** → Potential optic nerve/retinal disease.
2. **Standard Testing:**
 - **Snellen Chart:** Used at 6 m (20 ft).
 - **Rosenbaum Card:** Used at 36 cm (14 in) for near vision.
 - **Thresholds:** If < 6/240 (20/800), record as counting fingers, hand motions, light perception, or no light perception.

Pupillary Examination

1. **Basic Exam:** Test individually in dim light.
 - If light response is poor → test near response (to rule out Argyll Robertson, Parinaud's, or Adie's).
2. **RAPD Check:**
 - **Swing flashlight test** → identify Marcus Gunn pupil (indicates optic nerve disease).
3. **Anisocoria Evaluation:**
 - **Test with 0.125% pilocarpine** → identifies tonic pupil.
 - **Test with 1% apraclonidine** → dilates miotic pupil in Horner's syndrome.

Visual Field Mapping

1. **Localization Strategy:** Determine if lesion is pre-chiasmal, at the chiasm, or post-chiasmal.
2. **Pre-chiasmal (Monocular):**
 - **Central scotoma** → Macula/Papillomacular fibers.

- Arcuate scotoma → Optic nerve (Glaucoma, optic neuritis).
- Altitudinal field cut → Ischemic optic neuropathy.

3. Chiasmal:

- Bitemporal hemianopia → Symmetric compression of chiasm (e.g., Pituitary adenoma).
- Junctional scotoma → Asymmetric compression (e.g., Tuberculum sellae).

4. Post-chiasmal (Binocular):

- Homonymous hemianopia → Optic tract or radiations.
- Superior quadrantanopia → Meyer's loop (temporal lobe).
- Inferior quadrantanopia → Parietal lobe.

MANAGEMENT & TREATMENT

Acute Angle-Closure Glaucoma

1. Initial Treatment:

- Acetazolamide.
- Beta blockers.
- Pilocarpine.

1. Medical Therapy:

- Antiviral therapy.
 - Cycloplegics (to prevent scarring).
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KEY PEARLS & HIGH-YIELD POINTS

- **Pinhole Test:** Improvement → Refractive error; No improvement → Optic nerve disease.
- **PERRLA:** Redundant if light response is good; check near response if light is poor.
- **Horner's Syndrome:** Triad of miosis, ptosis, and anhidrosis (anhidrosis is inconsistent).
- **Roth's Spots:** Pathognomonic for subacute bacterial endocarditis; also seen in leukemia/diabetes.
- **Cherry-red Fovea:** Indicates central retinal artery occlusion (CRAO).
- **Amaurosis Fugax:** Transient ischemic attack of the retina → urgent stroke evaluation.
- **Legal Blindness:** 6/60 (20/200) or less in better eye OR binocular field $\leq 20^\circ$.
- **Optic Atrophy:** Not a specific diagnosis; reflects damage from various neuropathies.
- **Temporal vs. Nasal Pallor:** Temporal pallor is a more certain sign of optic atrophy than nasal rim pallor.