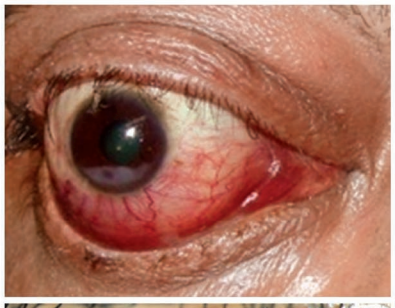

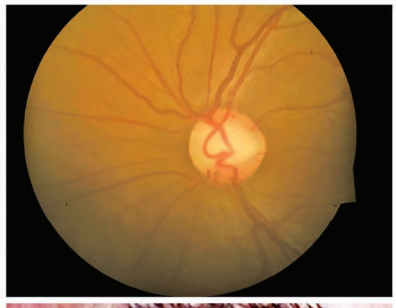

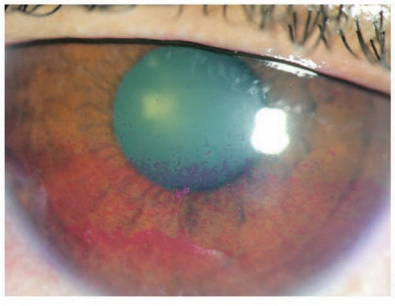

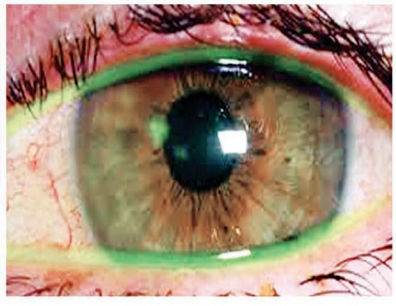

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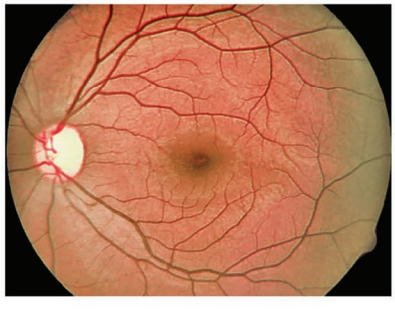

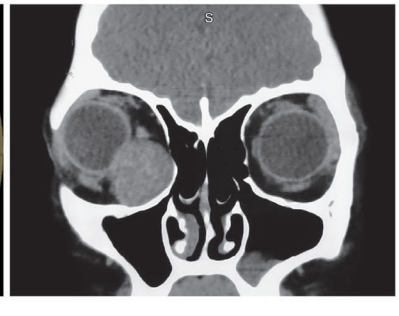

5th Edition

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Contents

1. Assessment of Visual Acuity 1

Rishabh Rathi, Siddharth Malaiya, Nitin Nema

- History *1*
- Principle *1*
- Development of Vision *1*
- Recording of Distant Visual Acuity *2*
- Macular Function Tests *10*

2. Clinical Refraction 15

Anuradha Narayanan, Swetha Saravanan, Nandhini Jaisankar

- Emmetropia *15*
- Ametropia *15*
- Diagnostic Procedures in Refractive Errors *17*

3. Color Vision and Color Blindness 24

Harinder Singh Sethi, Mayuresh Naik, Shweta Verma

- Color Vision *24*
- Factors Affecting Color Vision *24*
- Illumination *25*
- Theories of Color Vision *25*
- Anatomy of Color Vision *26*
- Anomalies of Color Vision *27*
- Color Vision Testing *29*
- New Digital and Automated Color Perception Test *39*
- Digital Color Vision Tests *39*
- Tablet-based Methods for Assessment of Color Vision Defect in Pediatric Population *41*
- Other Tests *45*

4. Slit Lamp Examination 52

Arulmozhi Varman

- History of Slit Lamp Development *52*
- Types of Illumination Used in Slit-lamp Examination *53*
- Gonioscopy *65*
- Slit-lamp Photography *68*

5. Vital Dyes and Stains in Ophthalmology 70

Pranessh Ravi, Shreesha Kumar Kodavoor

- Historical Preview *70*
- Biochemistry and Pharmacology of the Dyes *70*
- Dyes and Stains in Anterior Segment Diseases *71*
- Usage in Anterior Segment Diagnostics *71*
- Lissamine Green *76*
- Rose Bengal *77*
- Trypan Blue *77*
- Indocyanine Green *78*
- Brilliant Blue G *80*

6. Corneal Topography 84

Francisco Arnalich Montiel, Jorge L Alió Del Barrio, Jorge L Alió y Sanz

- Background *84*
- Videokeratoscopy *85*
- Fundamentals and Technological Approaches to Corneal Topography *85*
- Corneal Topography: Current Technologies *86*
- Performing a Good Topography Examination *89*
- Interpretation of Corneal Topography Maps *90*
- Quantitative Descriptors of Corneal Topography *94*
- Corneal Aberrometry: Fundamentals and Clinical Applications *98*
- Measuring Wavefront Aberration *99*
- Clinical Applications *104*
- Clinical Uses of Corneal Topography *105*
- Postoperative Cornea in Refractive Surgery *106*
- Postintrastromal Corneal Rings Implantation *110*
- Other Uses of Corneal Topography *112*

7. Corneal Tomography 114

Rohit Shetty, Ashwin Shetty, Luci Kaweri

- Pachymetry *114*
- Basic Principles of Tomography *115*
- Output *116*
- Topography *116*

- Specialized Maps *118*
- Screening for Ectasia Using Pentacam *124*
- Corneal Wavefront Analysis Using Pentacam *126*
- Contact Lens Fitting Using Pentacam *126*

8. Corneal Confocal Microscopy 128

Manotosh Ray, Chris HL Lim

- Optics *128*
- Confocal Microscopy of the Normal Cornea *129*
- Confocal Microscopy in Corneal Pathologies *132*
- Laser in Situ Keratomileusis *135*
- Infectious Keratitis *136*
- Corneal Grafts *138*
- Intracorneal Deposits *139*
- Vortex Keratopathy *139*

9. Diagnostic Procedures in Infectious Keratitis 141

Joveeta Joseph, Savitri Sharma

- In Vivo Corneal Confocal Microscopy *141*
- Laboratory Methods for Diagnosis of Nonviral Keratitis (Bacterial, Fungal, Parasitic) *142*
- Laboratory Methods for Viral Keratitis *157*

10. Tonometry 165

Rengaraj Venkatesh, Usha Tejaswini S, Indira Pegu

- Classification *165*
- Indentation Tonometers *165*
- Applanation Tonometers *167*
- Mackay-Marg Tonometer *169*
- Maklakov Applanation Tonometer *170*
- Noncontact Tonometer *170*
- Ocular-response Analyzer *171*
- Corvis ST Tonometers *171*
- Rebound Tonometry *171*
- Sterilization and Disinfection Protocols for Various Tonometers *172*

11. Gonioscopy 173

Ronnie Jacob George, A Parivadhini, Arun Narayanaswamy, L Vijaya

- Optical Principles *173*

12. Optic Disc Assessment in Glaucoma 185

Aparna Rao, Nikhil S Choudhari, G Chandra Sekhar

- Methods of Optic Disc Examination *185*
- Features of Glaucomatous Disc Damage *186*
- Why is this Important? *193*

13. Perimetry 195

Devendra Sood, NR Rangaraj

- Evolution of Octopus Perimeter *195*
- Basic Terminologies in Perimetry *197*
- Measurement Strategies *199*
- Humphrey Visual Field Analyzer *202*
- Humphrey Field Analyzer *211*
- Progression Analysis in Glaucoma *217*
- Recent Advances *218*

14. Imaging in Glaucoma 224

R Krishnadas, Niranjana Balasubramaniam

- Optical Coherence Tomography *224*
- Optical Coherence Tomography Angiography *234*
- Anterior Segment Optical Coherence Tomography *241*
- Ultrasound Biomicroscopy *242*

15. Diagnostic Approaches in Pupillary Disorders 245

Navin Jayakumar, Durga Priyadarshini S

Functional Anatomy and Physiology of the Pupil Reflexes *245*

- Pupil Light Reflex *245*

Dark Reflex *247*

- Sympathetic Outflow Pathway *247*
- Near Response *248*

Principles and Techniques of Pupil Examination *249*

- History *249*
- Pupil Size *249*
- Direct Light Reflex *249*
- Near Response *250*
- Pupil Dilation Reflex (Dark Reflex) *251*
- Ciliospinal Reflex *252*
- Hippus or Physiological Pupillary Unrest *252*
- Relative Afferent Pupillary Defect *252*
- Edge Light Pupil Cycle Time *256*

Disorders of Pupil Function *257*

- Afferent Pupillary Defects *257*
- Retinal Diseases *257*
- Asymmetric Retinal Adaptation With Induced Relative Afferent Pupillary Defect *257*
- Amblyopia *257*
- Media Opacities *257*
- Optic Tract Disorders *257*
- Pretectal Nucleus *257*
- Wernicke Pupil *258*
- Midbrain Disease *258*

- Paradoxical Pupil Constriction in the Dark *258*
- Efferent Pupillary Defects: Anisocoria *258*
- Pupil Size and Light Reflex *258*
- Anisocoria Greater in Darkness *259*
- Horner's Syndrome *259*

Anisocoria Greater in Light *262*

- Slit-lamp Examination *262*
- Pilocarpine Test *262*
- Causes of Anisocoria due to a Dilated Pupil *263*
- Tonic Pupil *263*
- Light-near Dissociation *264*
- Pupils in Traumatic Brain Injury *264*

16. Diagnostic Procedures in Uveitis 267

Jyotirmay Biswas, Reesha KR, Ranju Kharel (Sitaula), Janakiraman Palani

- Diagnostic Imaging Procedures *267*
- Invasive Procedures in Uveitis *274*
- Flow Cytometric Analysis *278*

17. A-scan Ultrasonography 283

Nazneen Nazm, Arup Chakrabarti, Meena Chakrabarti

- History *283*
- Physics of Ultrasound *283*
- Ultrasound Biometry Instrumentation *284*
- Examination Procedure *285*
- Technique for Measurement of Axial Length for Intraocular Lens Power Calculation *286*
- Difficult Situations in Biometry *288*
- Guidelines for Minimizing Errors *291*
- Biometry Through Different Intraocular Lens Materials *291*
- Immersion Vector A-/B-scan Biometry *292*
- Optimal Care of Biometry Equipment *292*
- Cleaning of Biometry Equipment *292*
- Calibration of Instrument *292*
- Intraocular Lens Power Calculation *293*
- Intraocular Lens Power Calculation in High-to-extreme Myopia *296*
- Toric Calculators *296*
- A-scan for Evaluation of Posterior Segment Diseases *297*

18. B-scan Ultrasonography 317

Anand Rajendran, Shruthi Suresh, Savithiri Palanivel

- Ocular Indications for Ultrasound Evaluation *317*
- Physics of Ultrasound *317*
- Ultrasound Probe or Transducer *318*
- Gain *318*
- Modes *319*
- Probe Orientations *319*
- Quantitative Echography *320*
- Kinetic Echography *321*
- Clinical Application of Ultrasound *321*
- Intraocular Tumors *326*
- Inflammatory Diseases of the Eye *327*
- Structural Anomalies of the Eye *328*
- Postsurgical Scenarios *329*
- Miscellaneous *330*

19. Ultrasound Biomicroscopy 333

Sushmita Kaushik, Savleen Kaur

- Apparatus *333*
- Scope *333*
- Methodology *333*
- Ultrasound Biomicroscopy in Glaucoma *335*
- Ultrasound Biomicroscopy in Strabismus *339*
- Ultrasound Biomicroscopy In Uveitis and Retinal Diseases *340*
- Ultrasound Biomicroscopy in Childhood Glaucoma *342*
- Ultrasound Biomicroscopy in Congenital Primary Aphakia *344*
- Ultrasound Biomicroscopy in Mass Lesion *346*
- Limitations of Ultrasound Biomicroscopy *346*

20. Intraocular Lens Power Calculation 349

Himanshu Shekhar, Vaibhav Namdev, Rajesh Sinha, Jeewan S Titiyal

- Intraocular Lens Power Formulas *349*
- Optical Biometry *354*
- Intraocular Lens Power Calculation in Special Situations *355*

21. Ophthalmoscopy 359

Pukhraj Rishi, Tarun Sharma

- Principles of Ophthalmoscopy *359*
- Indirect Ophthalmoscopy *359*
- Head-mounted Binocular Indirect Ophthalmoscopy *361*
- Monocular Indirect Ophthalmoscopy *365*
- Modified Monocular Indirect Ophthalmoscopy *366*
- Penlight Ophthalmoscopy *367*
- Direct Ophthalmoscopy *367*
- Hruby Lens Direct Ophthalmoscopy *368*
- Wide-angle Viewing System *369*

22. Fundus Autofluorescence 371

Anand Rajendran, Archana Rajamani, Sivaraman G

- Visualizing Fundus Autofluorescence *371*
- Assessing the Fundus Autofluorescence Image *373*
- Novel Autofluorescence *375*
- Clinical Applications *379*
- Classification of Fundus Autofluorescence Patterns in Early Age-related Macular Degeneration *382*
- Reticular Drusen *383*
- Detection and Quantification of Atrophy *383*
- Fundus Autofluorescence Findings in Early Choroidal Neovascularization *385*
- Fundus Autofluorescence Findings in Late Stage Choroidal Neovascularization *385*
- Fundus Autofluorescence Findings in Relation to Choroidal Neovascularization Classification *385*
- Optic Disk Drusen *386*
- Idiopathic Macular Telangiectasia *387*
- Chorioretinal Inflammatory Disorders *387*
- Acute Posterior Multifocal Placoid Pigment Epitheliopathy *388*
- Acute Syphilitic Posterior Placoid Chorioretinitis *388*
- Multifocal Choroiditis and Panuveitis *388*
- Multiple Evanescent White Dot Syndrome *389*
- Acute Zonal Occult Outer Retinopathy *389*
- Birdshot Chorioretinopathy *390*
- Pseudoautofluorescence *390*
- Ultrawidefield Autofluorescence *390*
- Future Directions and Limitations *390*

23. Fundus Fluorescein Angiography 393

Naresh Babu Kannan, Muthukrishnan Vallinayagam, Shivani Shrivastava

- Properties of Sodium Fluorescein *393*
- Principle of Fluorescein Angiography *393*
- Technique of Fluorescein Angiography *394*
- Side Effects and Complications *395*
- Normal Angiogram *396*
- Abnormalities in Fluorescein Angiogram *397*
- Fundus Fluorescein Angiography Findings in Specific Diseases *401*

24. Indocyanine Green Angiography..... 407

Rajesh Ramanjulu, P Mahesh Shanmugam, Pradeep Sagar

- History *407*
- Phases of Indocyanine Green Angiography *408*
- Adverse Reactions *408*
- Applications in Ophthalmology *408*
- Diagnostic Applications *408*
- Polypoidal Choroidal Vasculopathy *412*
- Central Serous Chorioretinopathy *413*
- Inflammatory Lesions of the Choroid *415*
- Tumors *415*
- Therapeutic Applications *416*

25. Multimodal Imaging in Posterior Segment Diseases..... 419

Giridhar Anantharaman, Kiran Chandra Kedarisetti

- Color Fundus Photography *419*
- Near-infrared Reflectance *420*
- Fundus Autofluorescence *420*
- Dye-based Angiography *422*
- Optical Coherence Tomography *422*
- Understanding Multimodal Imaging in Macular Diseases *424*

26. Anterior Segment Optical Coherence Tomography 439

R Ramakrishnan, Arijit Mitra, Madhavi Ramanatha Pillai

- Principle of Optical Coherence Tomography *439*
- Measurement Tools *440*
- Limitations *445*
- Applications *445*
- Swept Source Optical Coherence Tomography *456*
- Future of Anterior Segment Optical Coherence Tomography *458*

27. Posterior Segment Optical Coherence Tomography 461

Prapti Chheda, Rajeev R Pappuru

- Principles *461*
- Functions *461*
- Applications *462*
- Advantages of Optical Coherence Tomography *462*
- Advantages of Spectral Domain Optical Coherence Tomography *462*
- Analysis of Optical Coherence Tomography *462*
- Optical Coherence Tomography Biomarkers *470*

28. Optical Coherence Tomography Angiography in Retinal Diseases 477

Chaitra Jayadev, Rupal M Kathare, Rohit Agrawal, Santosh Gopi Krishna Gadde

- Principle *477*
- Optical Coherence Tomography Angiography of Retinal Disorders *479*

- Other Macular Neovascularizations *484*
- Polypoidal Choroidal Vasculopathy *485*
- Macular Telangiectasia Type 2 *486*
- Limitations of Optical Coherence Tomography Angiography *486*
- Wide-field Optical Coherence Tomography Angiography *489*
- Potential Clinical Applications of Deep Learning in OCTA *491*

29. Electrophysiological Tests 494

Subhadra Jalali, Srikanta Kumar Padhy, Nikitha Gurram Reddy

- Visual Electrophysiological Tests *494*
- Electrooculogram *495*
- Electroretinogram *498*

30. Diagnostic Procedures in Retinopathy of Prematurity 530

Yog Raj Sharma, Parijat Chandra, Rajvardhan Azad

- Retinopathy of Prematurity Classification *530*
- Retinopathy of Prematurity: Screening and Treatment *532*
- Widefield Retinal Screening *533*
- Fluorescein Angiography *534*
- Optical Coherence Tomography *535*
- Ultrasound Biomicroscopy *536*
- Ultrasonography *536*

31. Localization of Intraocular Foreign Body 539

Lingam Gopal

- Types of Intraocular Foreign Bodies *539*
- Evidence From Circumstances of Injury *539*
- Importance of a Foreign Body in the Management of an Injured Eye *540*
- Localization of Intraocular Foreign Body *540*
- Characterizing the FBS with Imaging *543*
- Intraoperative Localization *544*

32. Diagnostic Modalities in Intraocular Malignancies 547

Rolika Bansal, Ruju Unadkat, Santosh G Honavar

- Fundus Photography *547*
- Ultrasonography or Ultrasound Biomicroscopy *548*
- Fundus Fluorescein Angiography *548*
- Indocyanine Green Angiography *549*
- Optical Coherence Tomography and Optical Coherence Tomography Angiography *549*
- Computed Tomography *552*
- Magnetic Resonance Imaging *552*
- Fine Needle Aspiration Biopsy/Cytology *553*

33. Concomitant Strabismus: Diagnostic Methods 555

Digvijay Singh, Harinder Singh Sethi, Pradeep Sharma

- Features of Concomitant Strabismus *555*
- Terminology and Notation in Concomitant Strabismus *555*
- Diagnostic Methods *555*
- Qualitative Tests *557*
- Quantitative Tests *558*

34. Incomitant Strabismus: Diagnostic Procedures 567

Vijayalakshmi Perumalsamy, R Muralidhar

- Paralytic Strabismus *567*
- Restrictive Strabismus *581*

35. Diagnostic Procedures in Dry Eye Disease 585

Jayesh Vazirani, Devanshi Desai, Virender Singh Sangwan

- Definition *585*
- Dry Eye Diagnosis *585*
- General Physical Examination *585*
- Slit Lamp Examination *585*

36. Evaluation of Epiphora 591

AK Grover, Shaloo Bageja, Rituraj Baruah

- Applied Anatomy and Physiology of the Lacrimal Apparatus *591*
- Tear Secretion and Elimination *593*
- Evaluation of Epiphora *594*

37. Evaluation of Blepharoptosis 605

Shubhra Goel

- Applied Anatomy of Upper Eyelid *605*
- Classification *606*
- Evaluation of Ptosis *608*
- Ancillary Workup *613*
- Management *615*

38. Diagnostic Procedures in Proptosis 618

AK Grover, Shaloo Bageja, Amrita Sawhney

- Etiology *618*
- Pseudoproptosis *618*
- General Physical Examination *621*

- Ocular, Periorbital area, and Orbit Examination *621*
- Ophthalmic Investigations *625*
- Orbital Imaging *625*
- Systemic Investigations *631*

39. Orbital Imaging 634

Sumaiya Sehrish, D Ravi Varma

- Principles of Imaging Techniques *634*
- Applied Anatomy on CT Scan and MRI *642*
- Clinical Applications *647*

40. Videonystagmography 667

Mihir Kothari

- Videonystagmography System *667*
- Advantages of Videonystagmography Over Electronystagmography *667*
- Indications for Videonystagmography *668*
- How to Read a Videonystagmograph? *669*
- Clinical Examples *670*

41. Diagnostic Approaches in Ocular Motor Nerve Palsy 673

Shivcharan L Chandravanshi, Nitin Nema

- Oculomotor Nerve Palsy *673*
- Trochlear Nerve Palsy *676*
- Abducens Nerve Palsy *681*

42. Cerebral Angiography in Neuroophthalmology 686

Hima Pendharkar

- Indications for Cerebral Angiography *686*
- Contraindications *686*
- Preprocedural Workup *687*
- Consent *687*
- Angiography Machine *687*
- Anesthesia *687*
- Patient Position *687*
- Arterial Access *687*
- Procedure *687*
- Pediatric Patient *687*
- Complications *688*
- A Word of Caution *688*
- Postprocedure Care *688*
- Report *688*
- Advances in Cerebral Angiography: Three-dimensional Digital Subtraction Angiography *691*
- Advances in Arterial Access for Cerebral Angiography *691*
- Case Studies *691*

43. Diagnostic Procedures for Genetically Transmitted Eye Diseases 698

Syed Ali Fathima Afrin J, Srikrupa N Natarajan, Sarangapani Sripriya

- Specimens for NGS-based Molecular Diagnosis *698*
- Targeted Panel Sequencing *700*
- Whole Exome Sequencing *701*
- Whole Genome Sequencing *701*
- Mitochondrial Genome Sequencing *702*
- Third Generation Sequencing *702*
- Cytogenetic Abnormalities *702*
- Copy Number Variations *703*
- Accreditation for Genetic Testing Laboratories *705*
- Quality Management in a Genetic Laboratory *705*
- Variant Reporting *705*

Index 709

CHAPTER 15

Diagnostic Approaches in Pupillary Disorders

Navin Jayakumar, Durga Priyadarshini S

INTRODUCTION

The pupil controls retinal illumination to maximize visual perception. The dilator muscle causes pupil enlargement in dim illumination. This increases the amount of incoming light to supplement the retinal dark-adaptive mechanisms. In bright surroundings, the sphincter muscle can reduce retinal illumination by as much as 1.5 log units. The pupil also controls the depth of focus of the eye (by constriction) and reduces optical aberrations.

This chapter is divided into three parts in which relevant functional anatomy of the pupil, a step-by-step method of clinical examination of the pupil, and common disorders involving the pupil are described.

FUNCTIONAL ANATOMY AND PHYSIOLOGY OF THE PUPIL REFLEXES

PUPIL LIGHT REFLEX

The pupil light reflex consists of three components **(Fig. 15.1)**.[1]

1. The afferent arc originates in the eye and comprises the optic nerve, optic chiasm, and anterior two-thirds of the optic tract (the lateral geniculate nucleus does not play a role in the arc).
2. The interneuron in the midbrain is the pretectal olivary nucleus.
3. The efferent arc originates in the Edinger–Westphal subnucleus of the oculomotor nuclear complex, and continues along the oculomotor nerve to the ciliary ganglion, and then along the short ciliary nerves to the pupillary sphincter muscle.

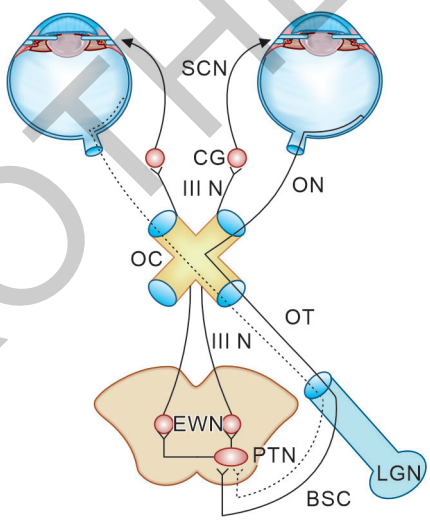


Fig. 15.1: The pupil light reflex pathway

(III N: oculomotor nerve; BSC: brachium of superior colliculus; CG: ciliary ganglion; EWN: Edinger–Westphal subnucleus of oculomotor nucleus complex; LGN: lateral geniculate nucleus; OC: optic chiasm; ON: optic nerve; OT: optic tract; PTN: pretectal nucleus; SCN: short ciliary nerves)

Afferent Arc

Photoreceptors

Both rods and cones contribute to the pupil light reflex. Under scotopic conditions, rods are far more sensitive to cones in eliciting a pupil contraction. In photopic conditions, the contribution of the cones to the pupil contraction is far more than rods. Pupil perimetry shows that the strength of pupil contraction is greatest when the central field is stimulated (where the photoreceptor and bipolar cell density is highest) and drops as the stimulus moves to the periphery.

Ganglion Cells

The retinal ganglion cells are the first neurons that give rise to action potentials that travel along their axons in the optic nerve and chiasm to cells in the lateral geniculate body and pretectal nuclei. Cells serving the afferent pupillomotor arc are most dense in the central retina. The major contribution to the pretectal olivary nucleus is from the γ ganglion cells (α- and β-cells project primarily to the lateral geniculate nucleus). The γ-cells have now been identified as the melanopsin-containing ganglion cells that play a role in other visually evoked reflexes such as the circadian rhythm.[2]

Melanopsin-containing ganglion cells are unique in that they can also be directly activated by light. Their photoreceptor-independent stimulation may explain why the pupil light reflex may be intact in patients with extensive photoreceptor damage or complete noncerebral monocular blindness. Studies comparing visual field defects with pupil perimetry (mapping of pupil sensitivity to light across the visual field) shows excellent correlation in certain diseases (e.g., anterior ischemic optic neuropathy) but large differences in other optic neuropathies (e.g., optic neuritis) where the visual field defect may be small but the defects in the "pupil field" are large.[3,4] These examples suggest that there may be relatively separate pathways for visual perception and pupillary response to light.

Interneuron

The pretectal olivary nucleus lies in the midbrain dorsolateral to the oculomotor nuclear complex. It is the interneuron of the pupil light reflex. It receives inputs from the pupillomotor ganglion cells of the ipsilateral temporal retina and the contralateral nasal retina (via the hemidecussation in the chiasm). These fibers leave the optic tract before its culmination in the lateral geniculate nucleus and enter the midbrain via the brachium of the superior colliculus.[5] The pupillomotor output from each pretectal olivary nucleus is distributed equally to the Edinger–Westphal nuclei of both sides to produce a pupil contraction of equal amplitude.

The contralateral homonymous hemifield organization of the afferent arc explains the finding of a contralateral relative afferent pupillary defect (RAPD) in an optic tract lesion.[6] This is because the intact ipsilateral temporal field (projecting along the uninvolved contralateral optic tract) provides greater pupillomotor input owing to larger retinal area being stimulated, than the intact nasal field of the contralateral eye.

Rarely, unilateral lesions of the brachium of the superior colliculus cause a "tectal RAPD" where pupillary response to stimulation of the contralateral hemifields is affected without contralateral homonymous hemianopia as would occur if the lesion were situated in the optic tract.[5,7]

Efferent Arc

Axons of cell bodies within the Edinger–Westphal subnucleus travel along the oculomotor nerve to synapse with the neurons in the ciliary ganglion in the orbit. The postganglionic fibers travel along with short ciliary nerves to innervate the sphincter muscle of the pupil.

Parasympathetic Oculomotor Nuclei

These subnuclei of the oculomotor nuclear complex contain cell bodies of preganglionic parasympathetic neurons that project to the ciliary ganglion and control pupil contraction to light and accommodation. Located in the dorsal midbrain they are the Edinger–Westphal nuclei, the anterior median nuclei, and the nucleus of Perlia. The Edinger–Westphal nuclei are responsible for the pupil contraction to light, while the anterior median nuclei are the primary neurons involved in producing accommodation response.

Oculomotor Nerve

The pupillomotor fibers are located superficially in the oculomotor nerve. This explains why the light reflex is "spared" in ischemic oculomotor nerve palsies, but "involved" in compressive etiologies such as aneurysms. The pupillomotor fibers are concentrated in the medial superior aspect of the oculomotor nerve as it leaves the brainstem and then gradually move medially and inferiorly as the nerve approaches the cavernous sinus.[8] These fibers are close to the posterior communicating artery. Aneurysms of this vessel compress the medial and superior aspect of the oculomotor nerve in the subarachnoid space causing a "pupil involved" oculomotor nerve palsy.

Ciliary Ganglion

The ciliary ganglion lies in the orbit 1.5–2 cm behind the globe between the optic nerve and the lateral rectus muscle, and in close association with the inferior division of the oculomotor nerve. The cell bodies are parasympathetic. Only 3% of the axons of these cell bodies project to the pupil sphincter, whereas over 94% reach the ciliary muscle.[9] This correlates with the relative masses of the two

muscles. Thus over 90% of the midbrain parasympathetic outflow is concerned with accommodation and not with pupil contraction.

Short Ciliary Nerves

Eight to twenty short ciliary nerves leave the ciliary ganglion in two to three bundles. Apart from postganglionic parasympathetic fibers destined for the ciliary muscle and pupil sphincter, these nerves also contain postganglionic sympathetic vasomotor fibers from the superior cervical ganglion and sensory afferents of the trigeminal nerve. The sympathetic and sensory roots to the ciliary ganglion do not synapse with its parasympathetic cell bodies. The short ciliary nerves enter the sclera at the posterior pole of the globe temporal to the optic nerve. They run anteriorly through the sclera and then in the suprachoroidal space.

DARK REFLEX

The pupil dilates in dim illumination and darkness. This reflex dilation is bilateral and follows a latency period that is slightly longer (about 300 ms) than the latency of the pupil light reflex. The dark reflex is mediated by the oculosympathetic pathway **(Fig. 15.2)**. The ocular sympathetic outflow originates at the hypothalamus and through a three-neuron pathway innervates the pupillary dilator muscle and vasomotor supply to the eye.

SYMPATHETIC OUTFLOW PATHWAY

First-order Neurons

The first-order neuron originates in the posterolateral hypothalamus. The axons descend in the brainstem and anterolateral columns of the spinal cord to synapse with the cell bodies of neurons of the ciliospinal center of Budge and Waller at C8–T2 spinal cord segments.

Second-order Neurons (Preganglionic)

The cell bodies of these preganglionic neurons lie in the ciliospinal center of Budge and Waller. Their axons exit the spinal cord via the ventral root to enter the paravertebral sympathetic chain. Here they pass without synapsing through the first thoracic (or stellate) ganglion close to the pleura at the apex of the lung. The fibers proceed

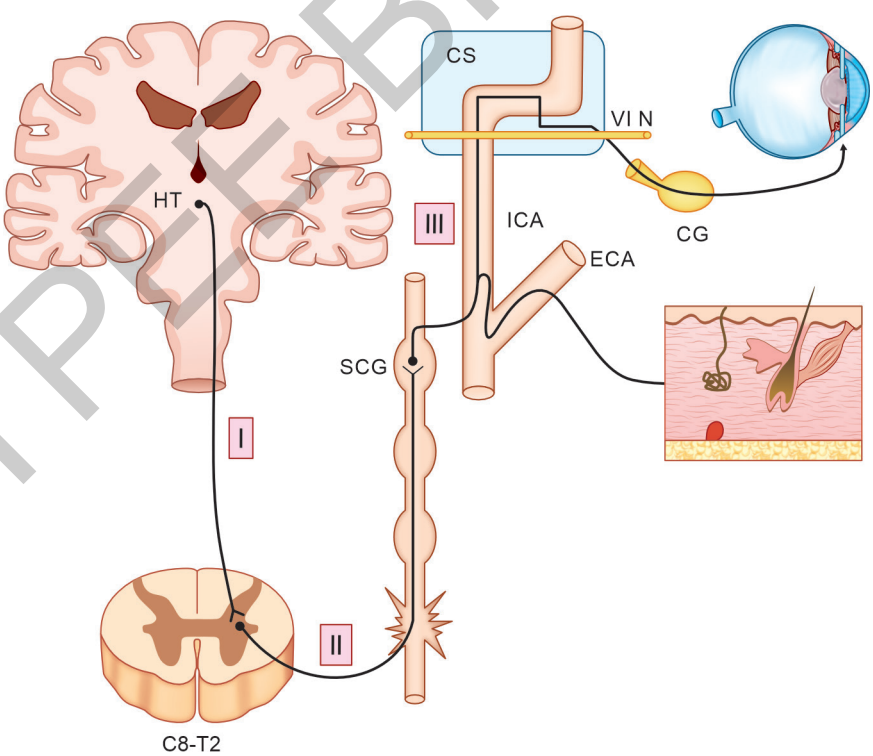


Fig. 15.2: The oculosympathetic pathway. I—first order neuron; C8-T2—spinal cord segments; II—second order neuron; III—third order neuron; VI N—abducens nerve
(CG: ciliary ganglion; CS: cavernous sinus; ECA: external carotid artery; HT: hypothalamus; ICA: internal carotid artery; SCG: superior cervical ganglion of the sympathetic chain)

uninterrupted through the inferior and middle cervical ganglia to terminate by synapsing with the postganglionic neurons in the superior cervical ganglion, which is located below the base of the skull between the internal jugular vein and internal carotid artery.

Two sets of branches exit from the superior cervical ganglion. The first communicates with C1 to C4 segments of the spinal cord and cranial nerves IX, X, and XII. The second set innervates the pharynx, the heart, and the sympathetic effectors in the head. This close relationship between the superior cervical ganglion and the lower cranial nerves explains their concurrent involvement in base of skull pathologies that produce a Horner's syndrome and lower cranial nerve palsies.

Third-order Neurons (Postganglionic)

The postganglionic fibers from the superior cervical ganglion accompany the internal carotid artery as a plexus surrounding the vessel. Almost immediately sudomotor and piloerector fibers destined for the face exit this plexus and follow the external carotid artery to the skin. The remaining fibers accompany the internal carotid artery intracranially through the foramen lacerum. In the cavernous sinus, the oculosympathetic fibers transiently fuse with the abducens nerve before joining the ophthalmic division of the trigeminal nerve.[10] The fibers then enter the orbit with the nasociliary nerve.

Besides the pupil dilator muscle, other parts of the eye also receive postganglionic sympathetic innervation. These include vasomotor fibers to the orbit, fibers to the branched melanocytes of the uveal tract, fibers to the extraocular muscles, the Muller's muscle, lacrimal gland, and the retinal pigment epithelium.

Innervation of the pupil dilator muscle occurs through the long ciliary nerves that bypass the ciliary ganglion. Some sympathetic fibers enter the ciliary ganglion, but exit it without synapsing along with the short ciliary nerves. The long and short ciliary nerves pass anteriorly and blend to form a rich plexus from which the iris muscles, ciliary muscle, and the vessel walls are supplied.

NEAR RESPONSE

When fixation shifts from far to near, three interrelated yet distinct responses are observed: (1) The pupils contract, (2) the lens accommodates (becomes more convex), and (3) the eyes converge. This triad is described as the near response **(Fig. 15.3)**. It is not described as a near reflex, because although the three occur virtually simultaneously,

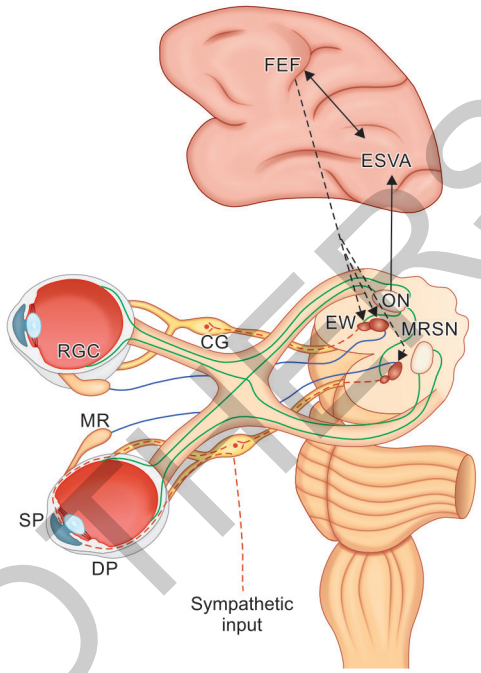


Fig. 15.3: Pupillary near reflex pathway: Near image blur—optic pathway—extra striate visual area—frontal eye field—(1) Edinger -Westphal nucleus—ciliary ganglion and sphincter pupillae—miosis and accommodation; (2) Medial rectus subnuclei—3rd nerve—medial rectus—convergence

(CG: ciliary ganglion; DP: dilator pupillae; ESVA: extra striate visual area; EW: Edinger -Westphal nucleus; FEF: frontal eye field; MR: medial rectus; MRSN: medial rectus subnucleus; ON: optic nerve; RGC: retinal ganglion cell; SP: sphincter pupillae)

it can be shown experimentally that each component can be selectively abolished or elicited without affecting the other two.[11]

The pupil light reflex and the near response miosis share a common final pathway from the Edinger–Westphal nucleus, along the oculomotor nerve, to the pupil sphincter muscle. The supranuclear pathways, however, are different. Visual stimuli (near-induced blur and temporal displacement of fixation on the retina) reach the visual cortex. From here, impulses travel to the frontal eye fields and thereon to the Edinger–Westphal nuclei in the midbrain (causing pupil constriction and lens accommodation) and to both medial recti subnuclei (producing convergence). In the Edinger–Westphal nucleus, the cells associated with the accommodation reflex are situated more ventral to those serving the pupil response to near. This arrangement explains how dorsal midbrain lesions produce "light-near dissociation" where the light reflex is impaired but the pupil constriction to near is uninvolved.

PRINCIPLES AND TECHNIQUES OF PUPIL EXAMINATION

HISTORY

Most patients with pupillary abnormalities are unaware of it. A useful way to evaluate the duration of an abnormality in pupil size or shape is to take a magnified look the pupils in photographs of the individual taken over time. This useful method is sometimes humorously described as "family album tomography" or "FAT scanning".

Symptoms associated with pupil size and shape abnormalities include photophobia, blurring of vision, and difficulty focusing when moving between environments with very different illumination levels. A large pupil causes blur in bright environments while a small pupil causes images to look relatively dim compared to the normal eye.

PUPIL SIZE

"Are the pupils equal in size? If not, is the difference greater in darkness or in light?" This is the first question to answer in the examination of the pupil. Pupil size can be examined by simple clinical examination using a pupil gauge. A pupil gauge is a simple handheld card (as a strip or a circle) that contains a series of solid circles or semicircles of diameters that increase in 0.5 mm steps. The gauge is placed adjacent to the pupil to estimate which circle (or semicircle) size best matches the patient's pupil size.

More sophisticated instruments such as a handheld pupil camera or infrared pupillography are also used.[12,13] An infrared pupillometer allows pupil observation even in complete darkness. The iris sphincter can be transilluminated to show areas of denervation or renervation.[14] Computer-based infrared video pupillography is increasingly used clinically to study pupil-related phenomena. They are used to measure pupil diameter in the planning of refractive surgical procedures, distinguish Horner's syndrome from physiological anisocoria, quantify RAPD, and for pupil perimetry.[15]

Slit-lamp biomicroscopy of the anterior segment also provides useful clues. Anterior uveitis (miotic pupil), corneal edema, and angle closure (mid-dilated oval pupil), iris transillumination, and areas of atrophy (post-trauma) are some examples. Segmental pupil contraction (Adie's pupil or oculomotor nerve palsy) can be visualized by turning off and on the slit-lamp beam. The magnification that a slit lamp provides can be used to detect a subtle RAPD. With the patient seated at a slit lamp and the slit illumination turned off, the swinging-flashlight test is performed using a transilluminator light, which is swung back and forth between the eyes. The direct pupil light response and the consensual light response can be observed under slit-lamp magnification.[16]

Pupil Size Measurement

Measure and record pupil size (in mm) in both eyes: (1) In bright illumination; (2) in dim illumination; and (3) for nearby using an accommodative target.

Use a scale or preferably a pupil gauge to determine pupil size. The illumination should be equal in both the eyes.

It is difficult to see the pupils in dim illumination, especially in a dark iris. For practical purposes, a "dim" torch or any minimal illumination is useful to visualize the pupil, for evaluation of pupil size in dim illumination in dark irides. The illumination must be just enough to allow visualization of the pupil edge.[17] Allow the illumination to fall evenly on both eyes at the same time, during measurement.

Anisocoria

Perceptible difference in pupil size between the two eyes is termed anisocoria: A difference of at least 0.4 mm between the two eyes. It is present in approximately 20% of young adults, with the prevalence increasing to 33% in the elderly.[18] Physiological anisocoria is asymptomatic and both pupils constrict briskly to bright light and near stimulation, and dilate briskly in dim illumination. Equality of responses seen on testing for reflexes is a good indicator of "normal" pupils whatever their size.

DIRECT LIGHT REFLEX

When bright light stimulates one eye, both pupils constrict. The constriction in the ipsilateral eye is called the direct reflex, while that in the opposite eye is called the consensual reflex. Provided there is no lesion in the autonomic pathways, the direct and consensual reflexes are clinically of equal magnitude.

Equipment

A good bright uniform light source is required. This is usually a pen torch or an indirect ophthalmoscope with the illumination turned to maximum. Clinically, the ideal instrument is an illuminator fitted to a direct ophthalmoscope handpiece. The light stimulus should be adequately bright. A very bright light [as seen in some

light-emitting diode (LED) torches] causes discomfort. It may produce spastic miosis lasting several seconds and will make determination of the normal light reflex difficult or impossible.[19] Blinking and tearing with glaring illumination make pupil evaluation difficult. A weak light source produces a correspondingly weak pupil constriction that can be misinterpreted as being sluggish, because pupil escape may occur under these conditions. Some torches have patchy uneven illumination. Do not use such torches with "central scotomas" as they may cause variable constriction of the pupils that can be misinterpreted.

Method

Keep room as dim as possible. Ask patient to look at a distant target (e.g., fixation target in mirror). Swing a bright light source in a quick motion onto the eye. Keep the eye illuminated for at least 3–4 seconds to observe events after the initial constriction. The direction of illumination should be from below and slightly temporal. This keeps the corneal images of the light source (first and second Purkinje images) outside the pupillary area, allowing a good visualization the entire pupil. Keeping a light source directly in front of the eye will stimulate the near reflex that will produce a pupil constriction. Observing pupil constriction by slit-lamp biomicroscopy is not the correct method of evaluating the direct reflex.

Observation

Record the two aspects of the reaction of the pupil: (1) Speed of reaction, "brisk or sluggish?" and (2) Sustaining of pupil constriction on continued illumination, "well-sustained or ill-sustained?".

The initial pupil constriction to bright light is called "pupil capture" (or phasic response).[20] Pupillary escape is an abnormal response where the pupil after initially constricting, redilates and returns to its original size. This may happen in patients with optic nerve or severe central or widespread retinal disease and lesions of the contralateral optic tract. Occasionally, pupillary escape occurs in a normal eye if a low intensity light source is used to test the direct reflex. The latency of the pupil reflex has been shown by pupillography to be 200–280 ms and a contraction that lasts 450 ms.[21]

Consensual Light Reflex

This test evaluates the efferent pathway of the pupil light reflex of an eye by stimulating the opposite eye with a bright light **(Fig. 15.4)**. The consensual reflex to light is clinically equal in velocity and amplitude as the direct reflex. However, it can be shown on infrared pupillography that the consensual reflex to light is in fact slightly less than that the direct reflex. This produces about 0.1 mm or less of what is described as alternating contraction anisocoria that is not normally visible on naked eye examination.[19]

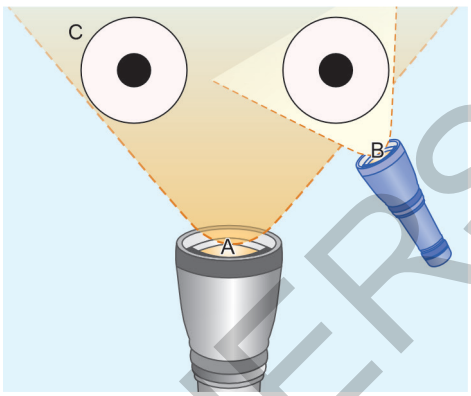


Fig. 15.4: The direct and consensual pupil reflex (A—dim diffuse light to illuminate both pupils equally for pupil visualization; B—bright light stimulus to elicit direct reflex in left eye; C—consensual reflex seen in right eye)

Method

Keep room as dim as possible. Ask the patient to look at a distant target (e.g., fixation target in mirror). Use a dim light source to illuminate the eye whose consensual pupil reflex is being evaluated. This light should be dim enough not to elicit a significant direct pupil reflex. Now swing a bright light source in a quick motion onto the opposite eye. As for the direct reflex record the speed and sustaining power of the pupil constriction.

NEAR RESPONSE

The pupil constriction to a near target is a synkinesis that also involves lens accommodation and convergence. A pupil with a brisk, well-sustained direct reflex will usually have a normal near reflex. Conversely, patients may present with an impaired light reflex with good near reflex suggestive of light-near dissociation. Light-near dissociation is characterized by a near response (tested in moderate light) that exceeds the best constriction that bright light can produce. Testing for the near reflex is therefore usually performed when the direct reflex is suspect. There is no clinical condition in which the light reflex is present and the near response is absent. A positive near reflex in a blind individual is an indication of the integrity of the efferent pathway and the pupil sphincter.

Equipment

The near response is best tested using a small accommodative target, which may sometimes include a Snellen optotype. A light source such as a pen torch should not be used. Also avoid using fingers as a near target.

Method

The ambient illumination should be moderate—sufficient enough for the patient to view the accommodative target. Ask the patient to keep his eyes focused on a distant target. Bring the near target about 30 cm in front of the patient's eyes and positioned between and slightly below them. Now instruct the patient to view the near target and observe the pupil constriction. Keeping the near target too low causes the eyes to look down, making it difficult to observe the pupils. Therefore, keep the target only slightly below the level of the eyes so as not to obstruct the patient's view of the distance target. The near reflex may occur slowly in certain situations (such as in the tonic pupil). To avoid missing this finding, keep observing the pupil for at least 10 seconds.

Sources of Error

Observe the degree of convergence as an indication of patient compliance—poor convergence indicates that the patient may not be trying enough to look at the near target, or that you are not holding the target near enough. This can result in an apparently poor near reflex.

Never judge near response by adding a near stimulus to a bright light stimulus. This action almost always produces additional constriction leading to "pseudo light-near dissociation".

Testing Near Response in a Blind Individual

Observe pupil constriction while instructing patient to voluntarily cross his eyes. This action does not require vision. Alternatively, look for the eye-closure pupil reaction. This is performed by instructing the patient to close his eyes while the examiner keeps the patient's lids open. This maneuver typically produces a strong pupil constriction.

PUPIL DILATION REFLEX (DARK REFLEX)

This test evaluates the function of the pupil dilator muscle. Normal pupil dilatation is a function of sphincter relaxation plus contraction of the dilator muscle. Pupil dilation normally occurs when ambient illumination changes from bright to dim. The pupils also dilate when fixation is shifted from near to distance. Very rarely, paradoxical pupillary responses may be observed in retinal diseases, where the pupil dilates in bright illumination and constricts to darkness.[22] Pupil dilation can also be elicited by sudden noise or when the back of the neck is pinched (ciliospinal reflex).

Method

Keep the ambient room illumination bright. Use minimal illumination evenly over both eyes just sufficient to keep both pupils visible in the dark. Switch off room illumination and observe dilatation of the pupils in the "dark".

Observation

Normally both pupils should dilate equally within 5 seconds and reach their widest size in 12–15 seconds. Note that the speed of the dilatation reflex is much slower than pupil constriction.

More importantly, look for a dilation lag indicating a weak dilator muscle. A dilation lag is an anisocoria developing after 2–4 seconds of dark and implies a sympathetic palsy (Horner's syndrome). Pupils that show dilation lag may take up to 25 seconds to return to their widest dilation with most of the dilation occurring about 10–12 seconds after onset of dark stimulus.

Alternative Methods

Another way to determine dilation lag is to take two sets of flash photographs of both eyes at 5 seconds (A) and 15 seconds (B) after onset of dark stimulus **(Fig. 15.5)**. If anisocoria (A) minus anisocoria (B) is >0.4 mm, then a Horner's syndrome is confirmed.[23]

Infrared videography of the pupils demonstrates dilation lag exquisitely even in very darkly pigmented pupils. Investigators have shown that redilation lag is the most sensitive diagnostic test (70% sensitivity and 95% specificity) to detect unilateral or bilateral Horner's syndrome.[24] The test involved videographing a dilated pupil in the dark that was then subjected to a bright light stimulus using a xenon arc flash. The time taken by the pupil from the peak of the light reflex to reach its baseline dilated state in the dark was measured. This is termed redilation and shows a delay in Horner's syndrome.

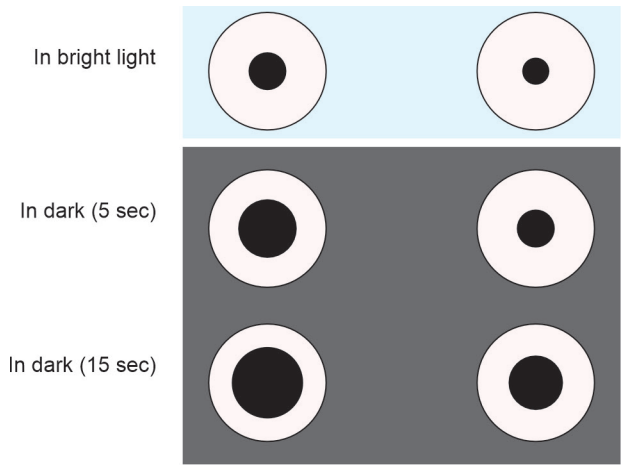


Fig. 15.5: Dilation lag in left Horner's syndrome. If anisocoria (at 5 seconds in dark) minus anisocoria (15 seconds after dark) is >0.4 mm, then a Horner's syndrome is confirmed

CILIOSPINAL REFLEX

The ciliospinal reflex induces bilateral pupillary dilation to painful stimuli applied to the skin of the neck (afferent C2 and C3). The reflex is mediated by second- and third-order sympathetic nerves to the pupil dilator muscle. Therefore, the ciliospinal reflex is a useful tool in differentiating central from peripheral sympathetic dysfunction.

Method

Pinch the side or back of the neck while observing both pupils.

Observation

Look for brisk bilateral simultaneous pupil dilation. Impaired dilatation reflex on one side suggests a lesion of the second- or third-order neurons of oculosympathetic pathway.

HIPPUS OR PHYSIOLOGICAL PUPILLARY UNREST

In ordinary illumination, healthy young pupils are in a state of almost constant motion. In the setting of prolonged illumination, both pupils constrict, then partly redilate, and begin to oscillate. In dim illumination, the pupils become larger and the oscillations are slower. In bright light, they become smaller and the oscillation rate increases. This physiological unrest is termed hippus and is present in all normal individuals. It is not triggered by accommodation. Hippus is reduced or absent in impairment of the afferent or efferent arc of the pupillary light reflex and in spastic miosis.

Oscillatory pupil movements occur with reduced wakefulness. These "fatigue waves" occur when a normal patient is very drowsy and is suppressed by mental activity. This phenomenon is similar but unrelated to hippus.

RELATIVE AFFERENT PUPILLARY DEFECT

In unilateral or asymmetrical bilateral optic neuropathy, the pupils exhibit differing velocities and amplitudes of constriction when testing the direct reflex. This difference is pupil reactivity, which is exploited by the swinging-flashlight test that exposes this difference as a RAPD.

Hirschberg proposed the concept of RAPD while describing a case of a young woman with unilateral retrobulbar neuritis. Gunn described his ability to differentiate retrobulbar neuritis from nonorganic visual loss by observing the differences in pupil light reflex between the eyes.[25] Kestenbaum popularized the concept of RAPD and credited Gunn with its discovery. Kestenbaum's test to demonstrate RAPD was modified by Levatin who proposed the "swinging-flashlight test" to demonstrate what was he labeled the Marcus-Gunn pupil (now described as RAPD).

Kestenbaum's Pseudoanisocoria Test

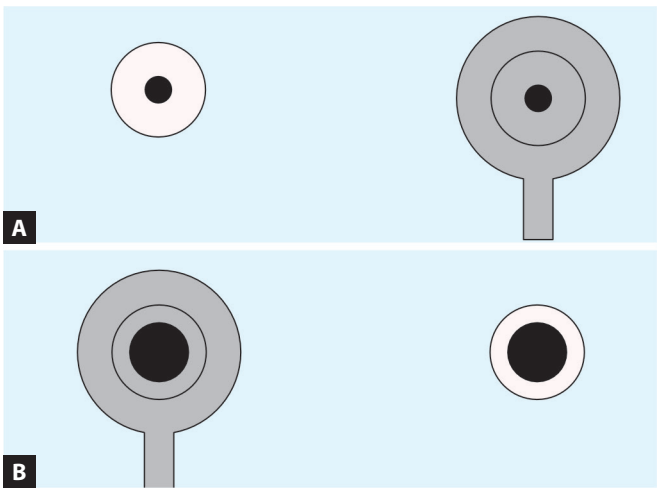
This test is not commonly performed nowadays being superseded by its refinement—the swinging-flashlight test. Nevertheless, it clearly demonstrates the underlying concept of the swinging-flashlight test (and can be used in the unlikely situation of a torch being unavailable!).

Method

With the patient in bright ambient illumination, measure the pupil size of one eye, keeping opposite eye occluded. Now perform the above maneuver with the initial eye occluded and measure the pupil size of the fellow eye **(Figs. 15.6A and B)**.[26]

Observation

Due to unequal afferent inputs in the presence of a unilateral (or asymmetrical bilateral) optic neuropathy, the affected pupil will be larger than the normal pupil. This is a pseudoanisocoria since the occluded and therefore, unobserved pupil is actually of the same size as the unoccluded pupil. The test will be positive in unilateral or grossly asymmetric bilateral optic nerve disease. It will not



Figs. 15.6A and B: Kestenbaum's pseudoanisocoria test in left optic neuropathy. Measure each pupil in bright ambient illumination keeping the opposite eye occluded. Right pupil (A) measures smaller than left pupil (B), indicating left afferent pupillary defect (note that this is a pseudoanisocoria since the occluded pupil is actually of the same size as the unoccluded pupil)

be positive in normals, and cannot be detected in patients with bilateral symmetric or subtle unilateral optic nerve disease.

Swinging-flashlight Test

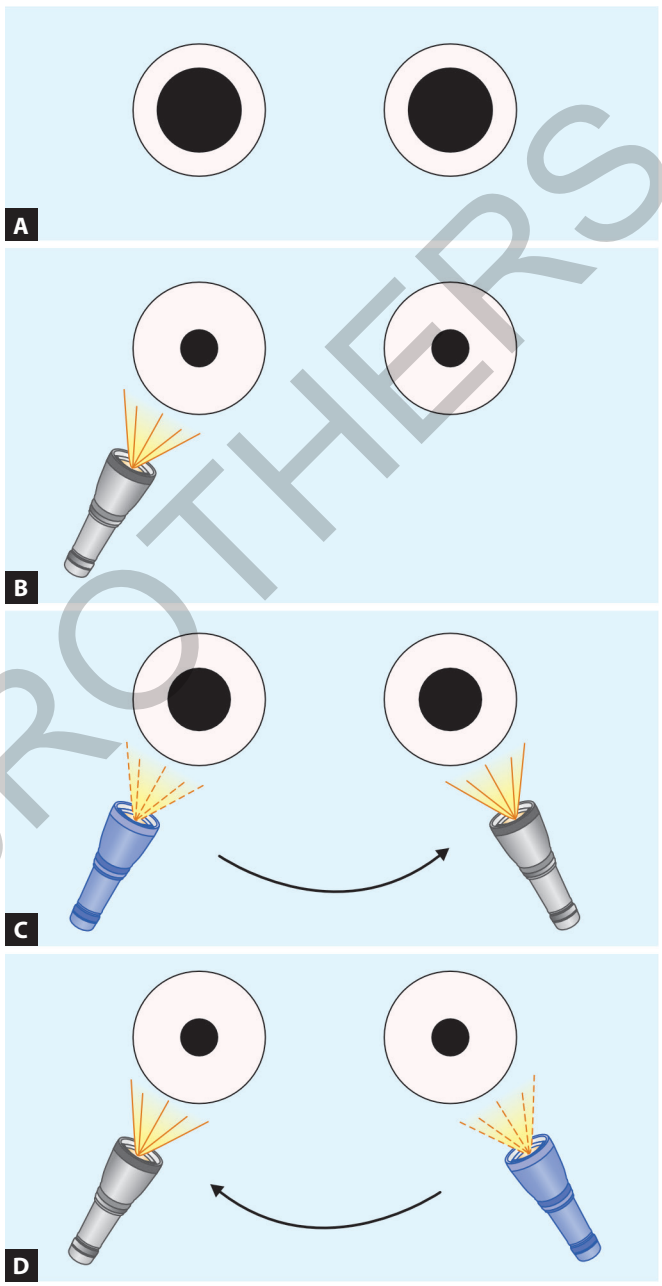
The test is named after Levatin. The pupil with the RAPD is described as a Marcus-Gunn pupil. The test compares optic nerve function between the two eyes **(Figs. 15.7A to D)**. It requires a normal efferent pathway (oculomotor nerve) and pupillary sphincter, on at least one side.[27]

Method

Keep ambient illumination as dim as possible. Ask the patient to look at a distant target (e.g., fixation target in mirror). Swing bright light source in a quick motion onto the eye and observe the reaction of the pupil. After about 3 seconds, rapidly swing the light source to the opposite pupil and observe its reaction. Continue swinging the light source in this manner between the two eyes to look for any relative sluggishness of the reflex or paradoxical dilation of the pupil of the illuminated eye.[26]

Observation

A paradoxical dilation of the pupil when light is directed on it signifies an optic nerve dysfunction in that eye. The pupil is called the Marcus-Gunn pupil indicating a RAPD in that eye.



Figs. 15.7A to D: Swinging-flashlight test. (A) Note equal pupil size in both eyes in the dark; (B) Stimulus on right eye produces visible right pupil constriction. Note that the unobserved left pupil constricts equally; (C) When the torch swings to the left eye, the left pupil paradoxically dilates. Note that the unobserved right pupil also "dilates" to the same size. Note also that compared to its size in the dark, the left pupil is actually constricting to light (compare left pupil size in A and C), though not to the same degree as the right pupil in (compare right pupil size between A and B); (D) Swinging the torch back to the right eye causes normal pupil constriction. Inference—left relative afferent pupillary defect

It usually takes less than a second to swing the light source from one eye to another. Too great a time interval between stimulation of the eyes results in pupil dilation in the intervening "dark period" causing a false negative test in eyes with mild optic nerve disease. Therefore, the "swing" between the two eyes should be reasonably rapid. It is recommended that the light source directly swings across the bridge of the nose as it passes from one eye to the other in a straight path. This avoids "false negatives".

Clinical Grading of Relative Afferent Pupillary Defect

Relative afferent pupillary defect can be clinically graded and has been correlated with corresponding values of neutral density (ND) filter log units **(Table 15.1)**.[28]

Quantifying Relative Afferent Pupillary Defect

Relative afferent pupillary defect is commonly quantified using ND filters of varying log units **(Figs. 15.8A to C)**.

A set of ND filters of 0.3, 0.6, 0.9, and 1.2 log units is adequate for clinical quantification.[29] These filters degrade the intensity of the stimulus in the normal eye. This simulates visual conditions of an eye whose optic nerve is affected. Using ND filters of appropriate log units over the normal eye, the afferent imbalance between the two eyes is neutralized. The RAPD ceases to exist and the Marcus-Gunn pupil disappears.

Method

Perform swinging-flashlight test to determine which eye has an RAPD. Place increasing log units of ND filters over the normal eye, while repeating the swinging-flashlight test. The end point is reached when the Marcus-Gunn pupil is abolished. The strength of the ND filter placed over the normal eye at the end point quantifies the degree of RAPD.

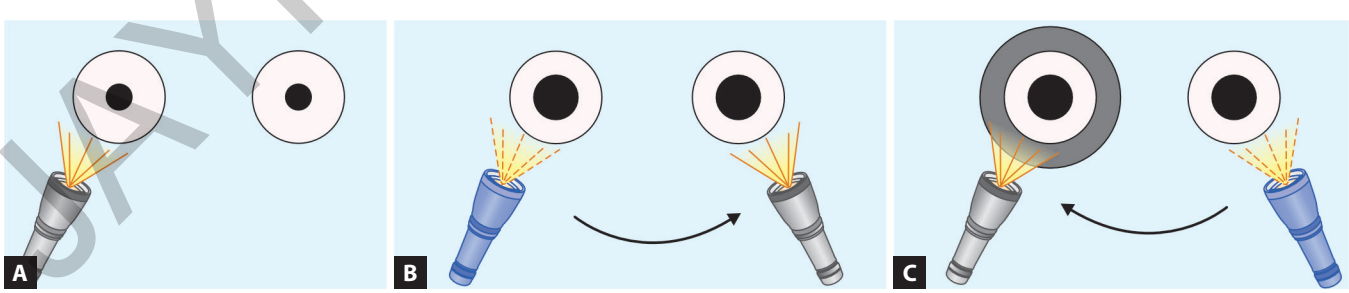
Cross polarizing filters have also been used instead of ND filters.[30] Their measurements correlate well with ND filter measurements. Cross polarizing filters showed exponential attenuation of light in contrast to linear attenuation by the ND filters.

A Bagolini or Sbisa bar has also been used to quantify RAPD. The Sbisa bar is used in amblyopia testing and to assess density of suppression in strabismus and consists of 17 sequential red filters. The Sbisa bar is a comparable

TABLE 15.1: Clinical grading of RAPD with corresponding values of ND filters

RAPD grade	*Features*	*Corresponding ND filter*
Grade I	Weak initial constriction and greater redilation	0.4
Grade II	Initial stall and greater redilation	0.7
Grade III	Immediate pupil dilation	1.1
Grade IV	Immediate pupil dilation following prolonged illumination of the good eye for 6 seconds	2.0
Grade V	Immediate pupil dilation with no secondary constriction	Infinity

(ND: neutral density; RAPD: relative afferent pupillary defect)



Figs. 15.8A to C: Quantifying RAPD with neutral density filters; (A) Bright light stimulus on the right eye produces visible right pupil constriction; (B) When the torch swings over to the left eye, the left pupil paradoxically dilated indicating a left RAPD; (C) An ND filter of appropriate strength reduces the intensity of light stimulus to the normal right eye creating an artificial optic nerve dysfunction (compare direct light reflex in right eye between A and C). Since both eyes are now equally "affected", swinging the torch back to the right eye causes no pupil constriction. The left RAPD has been neutralized

(ND: neutral density; RAPD: relative afferent pupillary defect)

instrument to the ND filter bar in measuring RAPD. Its availability in the clinical situation makes it a practical choice.[31]

Relative afferent pupillary defect can also be quantified by the swinging-flashlight test performed in combination with infrared video pupillography. With this method, 0.9 log units and above of RAPD can be reliably demonstrated.[32] Reliability is unsatisfactory for RAPD of <0.6 log units.

Interpretation of Relative Afferent Pupillary Defect

Relative afferent pupillary defect will be present in unilateral or asymmetric bilateral optic nerve disease. RAPD will not be present in normal, and cannot be detected in patients with bilateral symmetric optic nerve disease. In unilateral optic nerve disease, the degree of RAPD is broadly proportional to the severity of visual loss caused by optic nerve dysfunction. Although the presence of RAPD usually indicates optic nerve disease, trace RAPD may be observed in dense amblyopia and significant macular disease; in these situations, the degree of visual acuity impairment will be far greater than the degree of RAPD. Significant cataract in one eye causes the stimulating light to scatter and increase the pupillomotor "effectiveness" of the stimulus in that eye. This may induce an apparent RAPD in the opposite eye.

Swinging-flashlight Test with One Normal Pupil

Common situations where this might occur include sphincter damage (e.g., post intraocular surgery, acute angle-closure glaucoma), efferent pupillary defects (e.g., oculomotor nerve palsy with internal ophthalmoplegia), or poor visualization of the pupil (e.g., corneal opacity).

Testing in this situation is based on the principle of the consensual reflex **(Figs. 15.9A and B)**. The opposite pupil responds in an identical fashion to the pupil in which the light stimulus is applied.[19]

Method

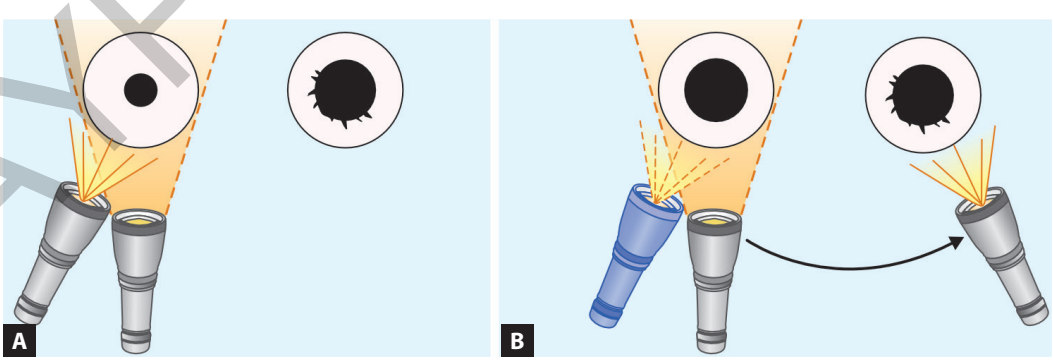
Use a dim light source with just sufficient illumination to illuminate the normal pupil. Perform the swinging-flashlight test as described earlier, by swinging the bright stimulating light source between the two eyes but observe only the dimly illuminated normal pupil.

Observation

If there is optic nerve disease in either eye, the observed pupil will exhibit an RAPD when the affected eye (irrespective of laterality) is stimulated. Remember the eye with RAPD is the eye being stimulated by the bright light source; this may or may not be the eye whose pupil is being observed.

Swinging-flashlight Test in Ocular Misalignment

With large angle strabismus or displacement of the globe in orbital disease, an off-axis stimulation of the squinting eye may result in a suboptimal pupil response and a false positive test in that eye. This is avoided by orienting the light source along the visual axis.



Figs. 15.9A and B: Testing for relative afferent pupillary defect (RAPD) with only one normal pupil in the right eye. The left pupil shows traumatic sphincter tears rendering it unreliable for pupil reflex evaluation. Dim illumination (shaded flashlight) is used to allow observation of the right pupil in the dark. (A) The right pupil shows a brisk direct reflex; (B) With the bright stimulus over the left eye, the left pupil is predictably immobile, but the right pupil dilates (by consensual reflex) indicating a left RAPD. Compare this response with the behavior of the right pupil in Figure 15.6C. The behavior is the same, the only difference being that in the present situation the right pupil (instead of the left) is being observed

Uncovering a Subtle Relative Afferent Pupillary Defect

The test is known as the "Tilt Test". This refinement of the swinging-flashlight test is useful when there is no visibly "detectable" RAPD, but *clinical suspicion of unilateral optic nerve dysfunction* exists.[33] This clinical prerequisite is significant as a small percentage of normals may also demonstrate small degrees of RAPD, which might also fluctuate between the eyes from time to time, a phenomenon described as an afferent equivalent of hippus.[34]

Equipment

The test requires a weak ND filter of 0.3 log units. A 0.3 log unit ND filter placed in front of an eye with a suspect RAPD heightens the inter-eye difference in pupillomotor drive to a detectable level, thus causing a Marcus-Gunn pupil to become apparent.

Method

Place 0.3 log unit ND filter in front of one eye and perform the swinging-flashlight test **(Fig. 15.10)**. Then repeat but with the same ND filter over fellow eye.

Observation

An eye with subtle optic nerve dysfunction will show a demonstrable RAPD in the test when the 0.3 log ND filter is placed in front of it. This occurs because the ND filter further reduces impulses along an already affected optic nerve. When the weak ND filter is placed over the normal eye, impulses along the normal optic nerve are slightly reduced. This would be approximately equal to the strength of impulses along the affected optic nerve in the fellow eye and thus no RAPD would be detected.

Clinical situation: 0.2 log unit clinically undetectable RAPD LE use 0.3 log unit ND filter for test			
0.3 log unit ND filter over	Total log unit defect		Observation
	RE	LE	
RE	0.3	0.2	No RAPD
LE	0.0	0.5 (0.3+0.2)	RAPD LE

Fig. 15.10: Tilt test for uncovering a subtle 0.2 log unit left RAPD that is clinically difficult to detect. A weak 0.3 log unit ND filter (depicted as a shaded circle) over the right eye induces a trace pseudoafferent defect rendering both eyes almost equal in terms of afferent input and no visible RAPD. However, transferring the ND filter to the left eye increases the afferent defect to 0.5 log unit (0.2 log unit afferent defect + 0.3 log unit ND filter = 0.5 log unit) rendering it clinically visible, confirming the presence of a subtle left afferent defect

(LE: left eye; ND: neutral density; RAPD: relative afferent pupillary defect; RE: right eye)

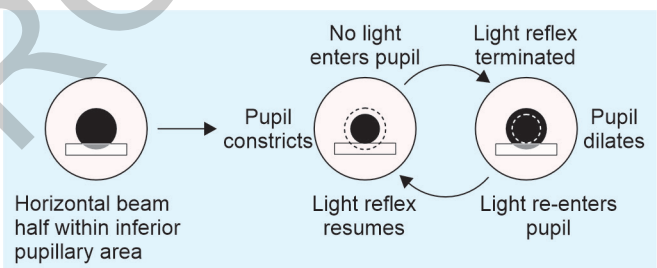


Fig. 15.11: Edge light pupil cycle time

EDGE LIGHT PUPIL CYCLE TIME

This test is not a particularly sensitive indicator of optic nerve disease when compared to the swinging-flashlight test. Nevertheless, it is useful when patients have bilateral symmetrical retinal or optic nerve dysfunction or have only one eye **(Fig. 15.11)**.[35]

Method

The patient is seated at slit lamp in dim ambient illumination and instructed to look at an imaginary distance target. Adjust the beam of the slit lamp to make it 0.5 mm thick and horizontally oriented. The beam is now focused on the inferior edge of the pupil such that half the thickness of the beam falls into the pupillary aperture. The light entering the pupil causes it to constrict. This constriction causes the entire slit beam to fall below the inferior pupil margin. The momentary loss of stimulus causes the pupil to partly dilate. The dilation now readmits half the slit beam into the pupil. This causes reconstriction of the pupil, and the cycle starts again.

Observation

Each constriction-dilation is a cycle. With a stop watch, measure the time for 30 cycles. Repeat the test for five 30-cycle rounds and calculate the time taken for a single cycle. Total number of cycles = 30 × 5 = 150. Edge light pupil cycle time (ELPCT) (in ms) = Total time (in seconds) for 5 rounds × 1,000/150. The normal range of ELPCT is 850–950 ms.

Diagnostic Procedures Explained in the Book

- Assessment of Visual Acuity
- Clinical Refraction
- Color Vision and Color Blindness
- Slit Lamp Examination
- Vital Dyes and Stains in Ophthalmology
- Corneal Topography
- Corneal Tomography
- Corneal Confocal Microscopy
- Diagnostic Procedures in Infectious Keratitis
- Tonometry
- Gonioscopy
- Optic Disc Assessment in Glaucoma
- Perimetry
- Imaging in Glaucoma
- Diagnostic Approaches in Pupillary Disorders
- Diagnostic Procedures in Uveitis
- A-scan Ultrasonography
- B-scan Ultrasonography
- Ultrasound Biomicroscopy
- Intraocular Lens Power Calculation
- Ophthalmoscopy
- Fundus Autofluorescence
- Fundus Fluorescein Angiography
- Indocyanine Green Angiography
- Multimodal Imaging in Posterior Segment Diseases
- Anterior Segment Optical Coherence Tomography
- Posterior Segment Optical Coherence Tomography
- Optical Coherence Tomography Angiography in Retinal Diseases
- Electrophysiological Tests
- Diagnostic Procedures in Retinopathy of Prematurity
- Localization of Intraocular Foreign Body
- Diagnostic Modalities in Intraocular Malignancies
- Concomitant Strabismus: Diagnostic Methods
- Incomitant Strabismus: Diagnostic Procedures
- Diagnostic Procedures in Dry Eye Disease
- Evaluation of Epiphora
- Evaluation of Blepharoptosis
- Diagnostic Procedures in Proptosis
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- Videonystagmography
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