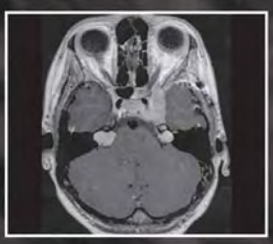
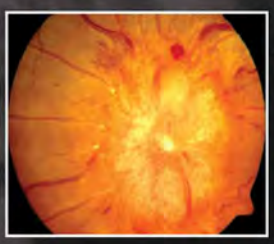
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Manual of Neuro-ophthalmology

2nd Edition

Editor
Amar Agarwal

Foreword
Nicole R Fram

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4

The Pupil

S Soundari, Amar Agarwal

NORMAL PUPIL

The pupil is an aperture present in the center of the iris and controls the amount of light entering the eye and reaching the retina. There is generally only one pupil but in some cases one can have more than one pupil and this condition is called polycoria.

PUPILLARY PATHWAYS

Introduction

When light is shone in one eye, both the pupils constrict.[1,2] Constriction of the pupil to which light is shone is called *direct light reflex* and that of the other pupil is called *consensual (indirect) light reflex*. If both pupils are illuminated simultaneously, the response summates. This means the constriction of each pupil is greater than the constriction noted when only one pupil is illuminated. Rods and cones initiate the light reflex.

Pupilloconstrictor Light Reflex Pathway

Afferent

The outer segments of the rods and cones are the receptors for both the visual pathway and the light reflex. When light is flashed on the eyes, the pupillary fibers run in the optic nerve. From there they cross in the optic chiasma and reach the optic tract (Fig. 4.1). From the optic tract they leave the visual pathway fibers (which continue to the lateral geniculate body) and reach the pretectal nucleus. This is an ill-defined collection of small cells anterior to the lateral margin of the superior colliculus. Internuncial fibers connect each pretectal nucleus with the Edinger-Westphal nucleus of both sides. Half of the postsynaptic fibers from the pretectal area curve around the periaqueductal gray matter to terminate in the ipsilateral Edinger-Westphal nucleus, while the other half cross, mainly via the posterior commissure to the contralateral Edinger-Westphal nucleus. This connection forms the basis of the consensual light reflex. Any given pretectal neuron behaves functionally as though it receives similar inputs from each eye and projects equally in each Edinger-Westphal nucleus.

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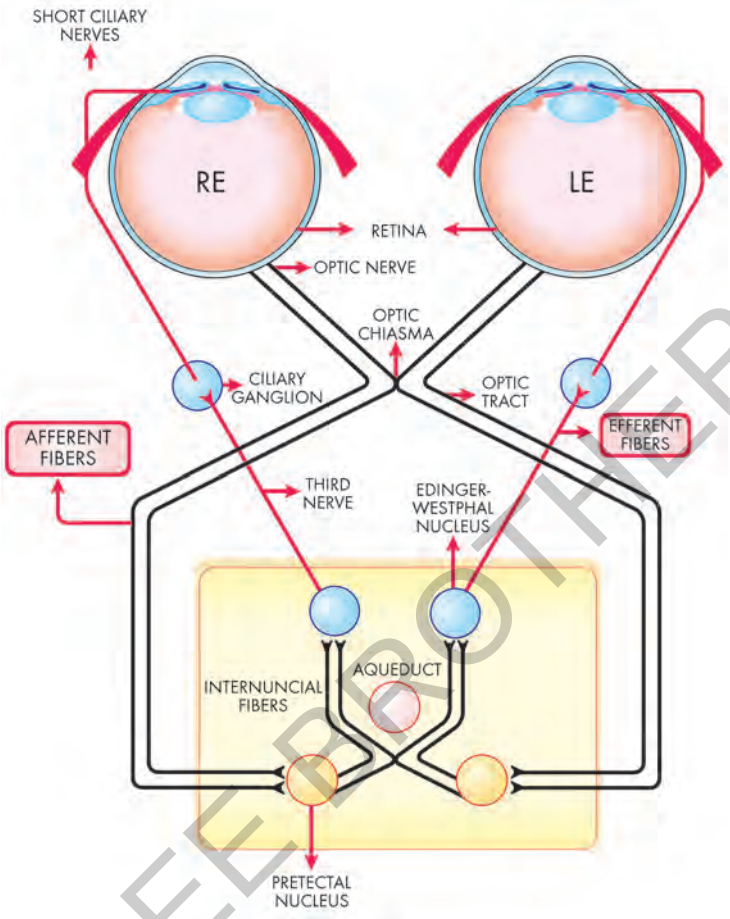


Figure 4.1 Pupilloconstrictor light reflex pathway

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Efferent

Preganglionic parasympathetic myelinated fibers now go to the ciliary ganglion via the third cranial nerve. They leave the III nerve in its branch to the inferior oblique. After reaching the ciliary ganglion they synapse there. Then postganglionic myelinated fibers pass through the short ciliary nerves to innervate the sphincter pupillae. Normally, postganglionic fibers are non-myelinated and this is the only exception to this rule. These are the parasympathetic fibers.

Sleep: Normally, there is a tonic inhibitory input from the cerebral cortex to the Edinger-Westphal nucleus and it is a diminution of this input that results in pupillary constriction during sleep.

Functions of the Light Reflex

- Pupillary constriction associated with light reflex protects against excessive bleaching of the visual pigments by reducing the amount of light entering the eye
- Light reflex helps in light and dark adaptation. This plays a role in maximizing visual acuity at different light levels. A large pupil will allow more light and so can allow greater acuity in dim illumination. On the other hand, a smaller pupil will limit the aberrations produced by the eye's refractive system and so can also allow greater acuity. It has been found that at any given light level, the size of the pupil strikes an optimum balance between these two factors.

Convergence near Reflex Pathway

Near reflex occurs on looking at a near object. It consists of two components—a convergence reflex and an accommodation reflex. The convergence reflex comprises convergence of the visual axes of the eyes with associated constriction of the pupil.

Afferent

The afferent starts from the medial recti. This travels centrally via the third nerve to the mesencephalic nucleus of the fifth nerve. From here the impulses travel to the convergence center in the tectal or pretectal region. Internuncial fibers from the convergence center then go to the Edinger-Westphal nucleus (Fig. 4.2).

Efferent

The efferent pathway is along the third nerve (similar to that of the light reflex). From the III nerve efferent fibers of convergence reflex relay in the accessory ciliary ganglion and from there reach the sphincter pupillae.

Accommodation Reflex Pathway

The accommodation reflex consists of increased accommodation and associated constriction of the pupil.

Afferent

The afferent starts from the retina. Impulses go from the rods and cones to the optic nerve, optic chiasma and optic tract (Fig. 4.3). Fibers then pass to the lateral geniculate body, optic radiations and striate cortex. The impulses reach area 19 of the occipital cortex. Internuncial fibers then pass from area 19 to the pontine center via the occipitomesencephalic tract. From the pontine center fibers pass to the Edinger-Westphal nucleus of both sides.

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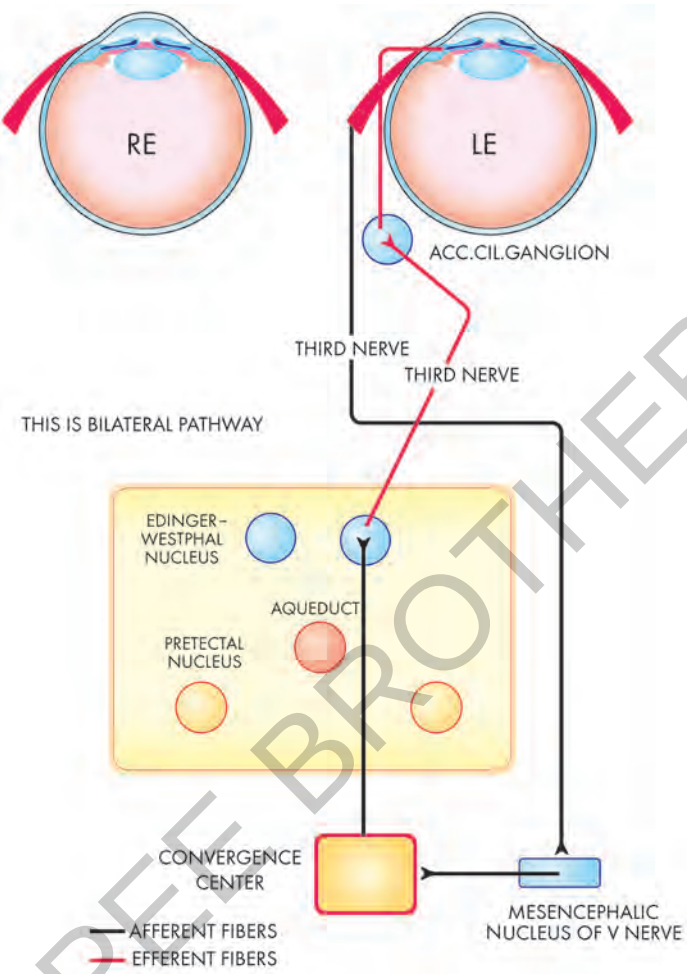


Figure 4.2 Convergence reflex pathway

Efferent

From the Edinger-Westphal nucleus the efferent impulses travel along the III cranial nerve and relay in the ciliary and accessory ciliary ganglion. From there, the fibers reach the sphincter pupillae.

Pupillary Dilatation Pathway

The pupillary dilatation starts from the posterior hypothalamus (Fig. 4.4). The pathway is the sympathetic pathway unlike the pupilloconstrictor pathway which is parasympathetic. From the hypothalamus first-order neurons start which reach the ciliospinal center of Budge. These fibers descend uncrossed. This center is located in the intermediolateral cell column of C_8, T_1 and T_2.

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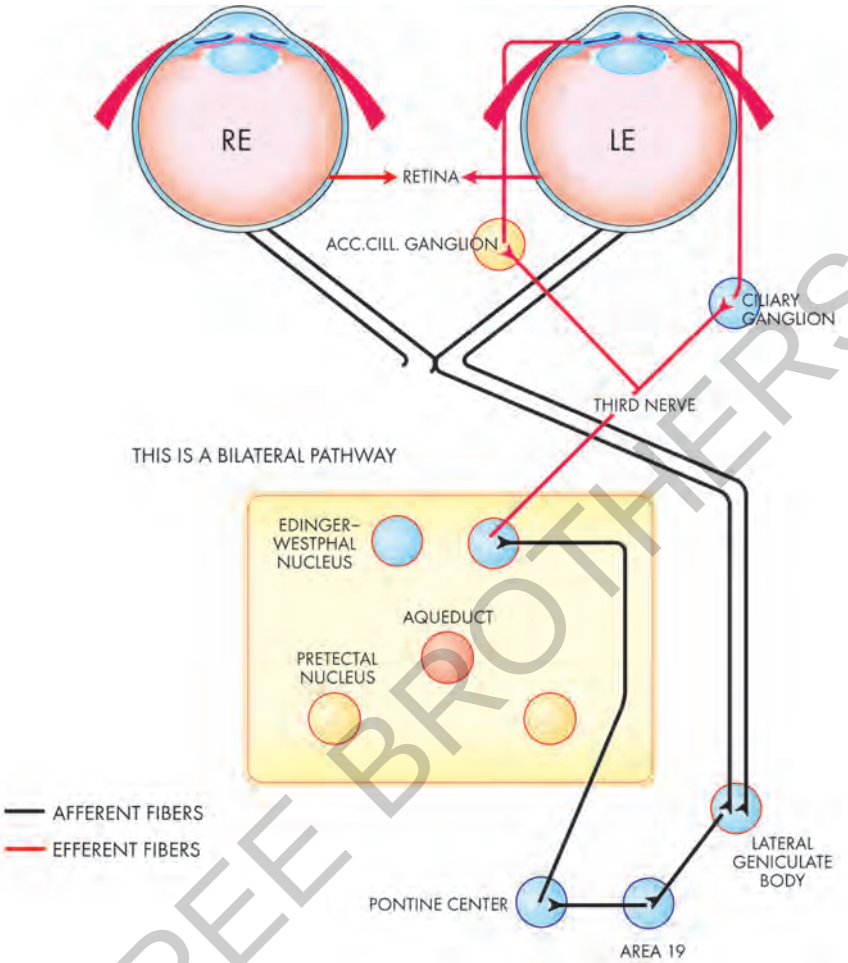


Figure 4.3 Accommodation reflex pathway

The second-order neuron starts from the ciliospinal center of Budge. These are the preganglionic fibers and they travel via the inferior and middle cervical ganglion to synapse in the superior cervical ganglion. During this long course, the preganglionic fibers are closely related to the apical pleura where they may be damaged by bronchial carcinoma. This is Pancoast's tumor. They can also be damaged during surgery on the neck.

The third-order neurons are postganglionic neurons which arise from the superior cervical ganglion. They ascend along the sympathetic plexus around the internal carotid artery to enter the skull. The fibers then join the sympathetic plexus around the ophthalmic artery. Then they go to the nasociliary nerve (branch of ophthalmic division of the trigeminal nerve) and pass through the ciliary ganglion. They do not synapse in the ciliary ganglion and reach the dilator pupillae via the long ciliary nerves.

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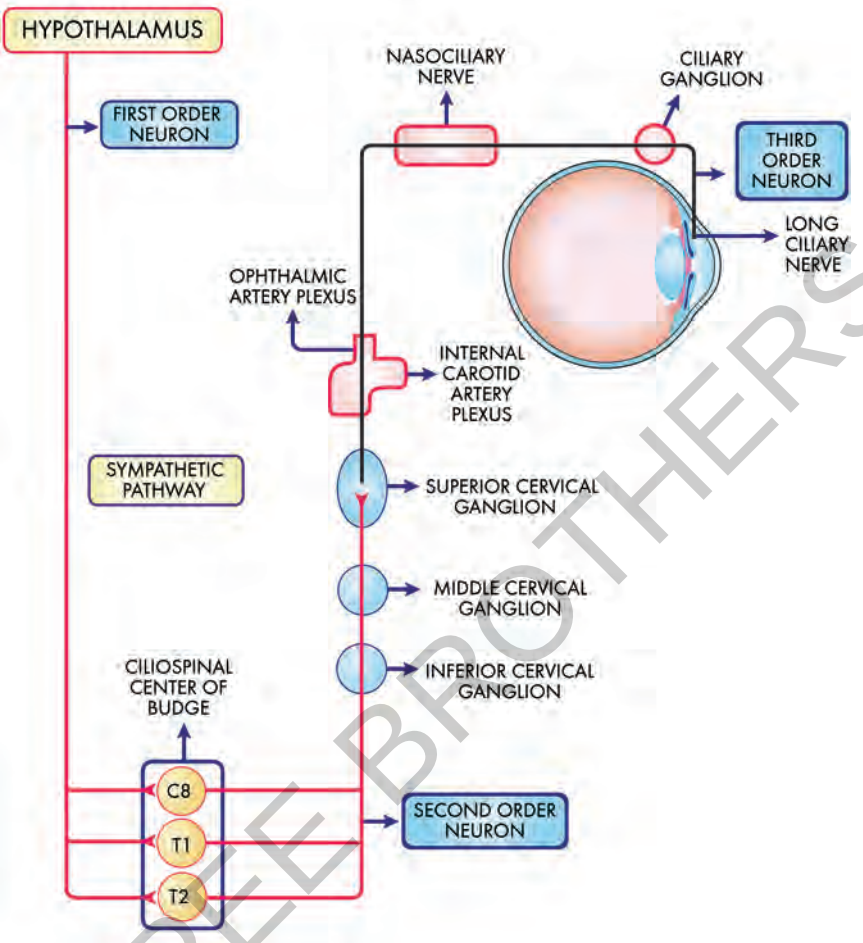


Figure 4.4 Pupillary dilatation pathway

Darkness Reflex

When a person goes from a lighted environment to darkness, the pupils dilate. This dilatation has two causes—the first is simply abolition of light reflex with consequent relaxation of the sphincter pupillae, and the second is contraction of the dilator pupillae supplied by the sympathetic nervous system. The pathways involved in dark reflex are presumably the same as those of light reflex, since the dilatation at the end of long light stimulation also involves both relaxation of the sphincter pupillae and contraction of the dilator pupillae.

Psychosensory Reflex

Psychosensory reflex refers to dilatation of the pupil in response to sensory and psychic stimuli. A large number of differently described reflexes fall into this

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category. They are not seen in a newborn baby, but appear in the first few days of life, developing fully at the age of 6 months. Their mechanism is very complex and their pathways have still not been elucidated. It is believed that the mechanism of psychosensory reflexes is a cortical one and the pupillary dilatation results from two components—a sympathetic discharge to the dilator pupillae and an inhibition of the parasympathetic discharge to the sphincter pupillae.

PUPIL CYCLE TIME

A small beam of light focussed at the pupillary margin induces regular, persistent oscillations of the pupil. These oscillations can be timed with a stopwatch. The period of the average complete cycle is called the *pupil cycle time*. It is prolonged in optic neuritis and in compressive optic neuropathy.

LESIONS OF THE PUPIL

There are various lesions, which can occur in the pupil (Fig. 4.5) depending upon the location of the lesion. These are as follows:

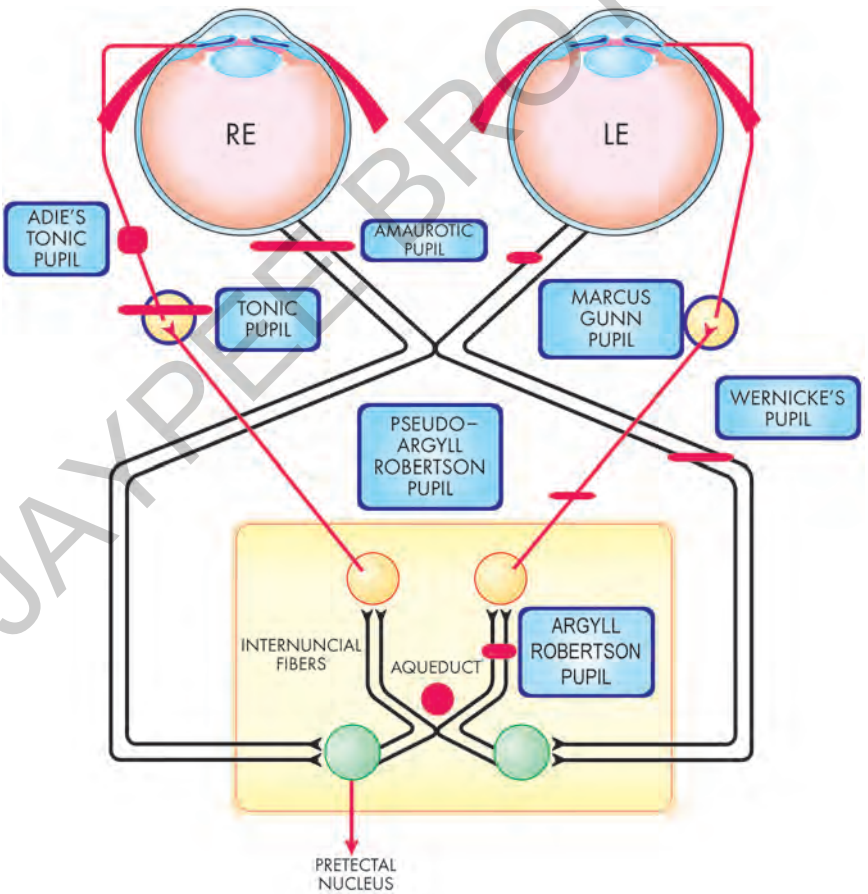


Figure 4.5 Lesions of the pupil

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Amaurotic Pupil

Amaurotic pupil is a total afferent pupillary defect (Fig. 4.5). A complete optic nerve or retinal lesion leading to total blindness on the affected side causes it. The eye has no perception of light. The points in an amaurotic pupil are:

- The pupil neither reacts to direct light stimulation, nor does it create a consensual light reflex in the opposite eye
- When light is shone on the opposite eye, there is a good light reflex in that eye and a good consensual light reflex in the affected eye
- This shows that the defect is an afferent defect and the efferent system is normal.

Marcus Gunn Pupil

Swinging Flashlight Test

A Marcus Gunn pupil is a relative afferent pupillary defect. An incomplete optic nerve lesion or severe retinal disease causes it. It is best tested by the swinging flash-light test. To perform this test (Fig. 4.6), a bright flashlight is shone onto one pupil and constriction is noted. Then the flashlight is quickly moved to the contralateral pupil and the response is noted. This swinging to and fro of the flashlight is repeated several times while the pupillary response is observed. Normally, both pupils constrict equally and the pupil to whom light is transferred remains tightly constricted. In the presence of a relative afferent pupillary defect in one eye, the affected pupil will dilate when the flashlight is moved from the normal eye to the abnormal eye. This is a paradoxical response. This is called the Marcus Gunn pupil and is the earliest indicator of optic nerve disease even in the presence of a normal visual acuity.

Grades

Marcus Gunn pupil can be graded depending on the response to the swinging flashlight test. During the swinging flashlight test, if the amount of light information transmitted from one eye is less than that carried from the fellow eye, the following phenomenon may be noted when the light is swung from the normal eye to the defective eye. Thus, Marcus Gunn pupils can be graded.

- 3–4 + Marcus Gunn pupil: There is immediate dilatation of the pupil, instead of normal initial constriction
- 1–2 + Marcus Gunn pupil: No change in pupil size, initially, followed by dilatation of the pupils
- Trace Marcus Gunn pupil: Initial constriction, but greater escape to a larger intermediate size than when the light is swung back to the normal eye.

Lesion

Marcus Gunn pupil sign indicates an asymmetry of conduction and will not be present in symmetric bilateral lesions of the optic nerve or retinal disease.

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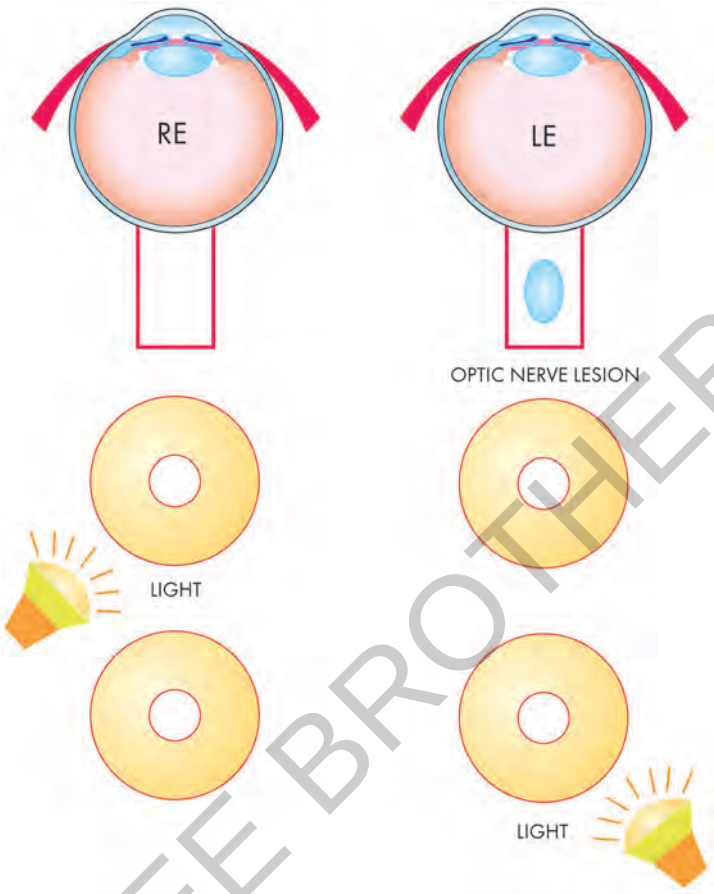


Figure 4.6 Swinging flashlight test

A Marcus Gunn pupil will not occur if the lesion is in the optic chiasma or optic tract as in these areas fibers are present from the opposite eye also.

Important Points

- There is no such thing as bilateral Marcus Gunn pupil. There may be bilaterally reduced direct response of the pupils to light, resulting in light-near dissociation, but the Marcus Gunn phenomenon requires asymmetry of the afferent light transmission
- Opacities of the ocular media (corneal scar, cataracts or vitreous hemorrhage) will not cause a Marcus Gunn pupillary phenomenon if a strong enough flashlight is used
- Extensive retinal damage will cause a significant Marcus Gunn phenomenon.

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Wernicke's Hemianopic Pupil

Wernicke's hemianopic pupil indicates the lesion in the optic tract (Fig. 4.5). In this condition, light reflex is absent when light is thrown on the temporal half of the retina of the affected side and nasal half of the retina of the opposite side. Light reflex is present when the light is thrown on the nasal half of the affected side and temporal half of the opposite side. The patient also has homonymous hemianopia as the lesion is in the optic tract.

Argyll Robertson Pupil

Lesion

A lesion (neurosyphilis) causes it in the region of tectum (Fig. 4.5). The lesion is in the region of the sylvian aqueduct when the fibers from the pretectal nucleus go to the Edinger-Westphal nucleus. An Argyll Robertson pupil occurs when in addition to involvement of the internuncial neurons, there is a disturbance of the normal inhibitory pathways from the reticular activating system upon the parasympathetic Edinger-Westphal subnucleus. The result of this inhibition is excessive parasympathetic activity and small pupils.

Features

- The vision in the affected eye is normal
- There is no reaction to light
- The near reflex is normal and the pupils react as the convergence and accommodation reflex fibers are not affected. The fibers that mediate the pupillary near response lie ventral to the internuncial neurons that enter the Edinger-Westphal nucleus
- The pupils are miotic and irregular
- The pupils dilate very poorly with mydriatics.

Light-near Dissociation

There are many causes of light-near dissociation. They are shown in Figure 4.5. They are:

- Argyll Robertson pupil
- Bilateral complete afferent pupillary defect as in bilateral optic atrophy. In these, the convergence reflex pathway (Fig. 4.2) is not affected as it starts from the medial recti, so there is a light-near dissociation.
- Lesions in the pretectal area as once again the near reflex are not affected. This occurs in Parinaud's syndrome.

Pseudo-Argyll Robertson Pupil

In this, there is third nerve palsy with aberrant regeneration of medial rectus innervation into the sphincter innervation pathway.

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How to Test for a Pupillary Near Response

The near response should be tested in good room light, so that the patient's pupils are midsized and the near object is clearly visible. The patient is given an accommodative target to look at. Watching for convergence helps the examiner judge how hard the patient is trying.

Tonic Pupil

Damage to the ciliary ganglion or short ciliary nerves (Fig. 4.5) produces the tonic pupil. The features are:

- Reaction to light is absent and to near reflex very slow and tonic
- Accommodative paresis is present
- The affected pupil is larger
- It is generally unilateral
- Cholinergic supersensitivity of the denervated muscle occurs. The normal pupil does not constrict with 0.125 percent pilocarpine whereas the tonic pupil does.

It could be due to viral infections affecting the ciliary ganglion like herpes zoster. It could also be due to diabetes or alcoholism.

When a tonic pupil is associated with absent deep tendon reflexes in the lower extremities the condition is called *Adie's tonic pupil*. The lesion is due to denervation of the postganglionic supply of the sphincter pupillae and ciliary muscle of unknown etiology. It generally occurs in women.

The near response of the pupil generally exceeds the light reaction in Adie's syndrome. The near response is slow and steady and on looking back into the distance it tends to hold the contraction for a few seconds. Thus, it is *tonic*. The reasons for this behavior are not very well understood. The slowness of the tonic pupil might be due to the diffusion of acetylcholine through the aqueous to the supersensitive receptors of the iris sphincter. The near reaction is not spared in Adie's syndrome—it is restored. Aberrant regeneration of fibers occurs that were originally destined for the ciliary muscle into the iris sphincter. So with every effort to focus the eye on a near object, impulses spill into the sphincter, constricting the pupil. Accommodative fibers in the short ciliary nerves outnumber sphincter fibers by 30 to 1. This means that the ciliary muscle will probably receive appropriate reinnervation, but the odds against the iris sphincter receiving the right fibers are very high. Thus, with random regeneration of fibers the power of accommodation is likely to recover, whereas the light reaction will not. At the same time, the sphincter is likely to be served by aberrant accommodative impulses that constrict the pupil firmly with every near effort.

Hutchinson's Pupil

Hutchinson's pupil occurs in comatose patients with unilaterally dilated, poorly reactive pupils. It is due to ipsilateral, expanding intracranial supratentorial mass (tumor or subdural hematoma) that is causing downward displacement of the hippocampal gyrus and uncal herniation across the tentorial edge with entrapment of the third nerve. The pupillomotor fibers travel in the peripheral portion of the third nerve and are subject to early damage from compression.

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This abnormality typically heralds the onset of a III nerve paralysis and is an internal ophthalmoplegia. The appearance of an internal ophthalmoplegia in a patient with a suspected or proven supratentorial mass is an ominous sign and indicates urgent surgical intervention.

Horner's Syndrome

Introduction

In Horner's syndrome, there is a lesion of the sympathetic system. There can be three types—central, preganglionic or postganglionic (Fig. 4.7). The characteristic features are as follows:

Ptosis: There is mild to moderate ptosis due to paralysis of the Muller's muscle which is supplied by the sympathetic system.

Enophthalmos: There can be upside down ptosis also and this is due to weakness of the inferior tarsal muscle. In this, there is elevation of the inferior eyelid. This leads to an apparent *enophthalmos*.

Miosis: There is moderate miosis due to unopposed action of the sphincter pupillae following paralysis of the dilator pathway. Pupillary reactions are normal to light and near. When the lights are turned off the Horner's pupil dilates more slowly than the normal pupil because it lacks the pull of the dilator pupillae. This is called *dilatation lag*.

Facial anhydrosis: Reduced sweating on the ipsilateral face and neck occurs. This is characteristic of preganglionic Horner's syndrome.

Heterochromia iridis: When the sympathetic ocular innervation is interrupted early in life (congenital Horner's syndrome) the pigment of the iris stroma fails to develop producing heterochromia iridis.

Central Horner's Syndrome

This occurs in lesions located between the hypothalamus and the ciliospinal center of Budge (Fig. 4.7). It is due to brainstem vascular lesions or demyelinating lesions. The patient will also have brainstem signs and sudden onset of vertigo.

Preganglionic Horner's Syndrome

In this, the lesion is located from C_8 to T_2 and the superior cervical ganglion. In other words, the second-order neurons are affected. This can occur in Pancoast's tumor of the lung or surgery in the neck. There is anhidrosis of the face and neck as the fibers for sweating come out from the superior cervical ganglion.

Postganglionic Horner's Syndrome

The lesion involves the third-order neurons from the superior cervical ganglion to the dilator pupillae. It can occur in cavernous sinus lesions or head trauma. There is ipsilateral vascular headache.

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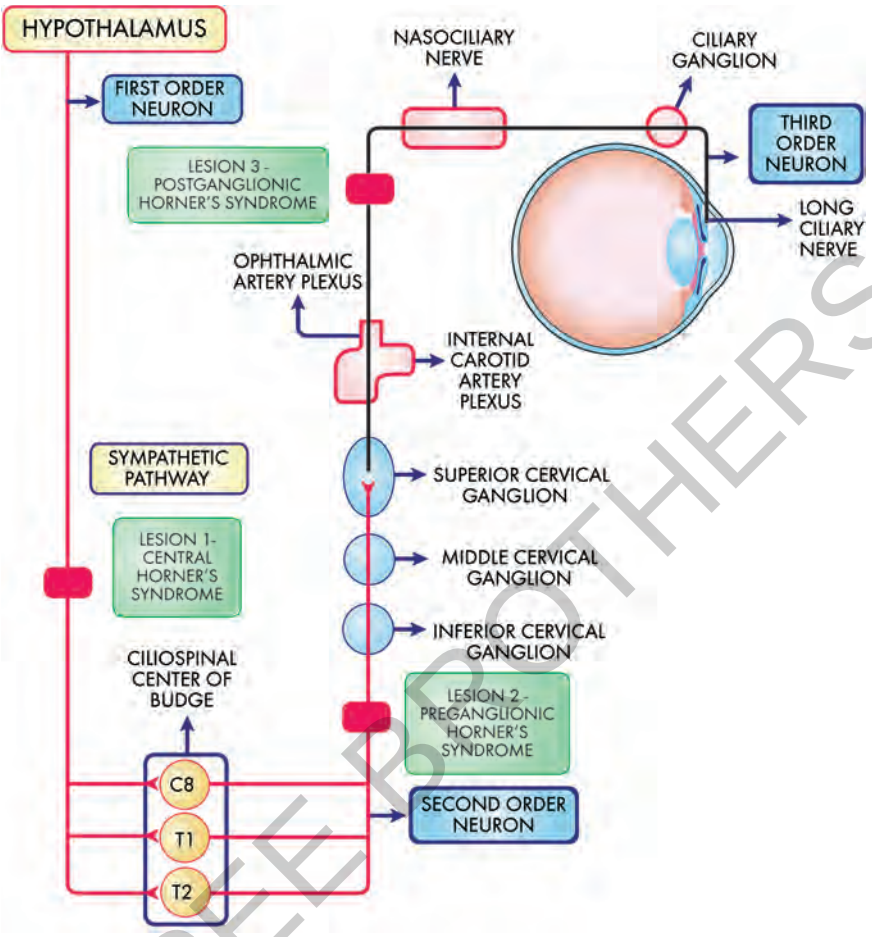


Figure 4.7 Horner's syndrome

Pharmacological Tests for Horner's Syndrome

Cocaine test: When 4 percent cocaine is instilled in both eyes, the normal pupil will dilate, but the Horner's pupil will not (Fig. 4.8). All Horner's pupils, no matter where the defect in the pathway is located, will dilate poorly to cocaine. Thus, cocaine helps in establishing the diagnosis of sympathetic denervation and not in localizing the site of lesion.

Hydroxyamphetamine test: When 10 percent drops of this drug are instilled into both eyes, in a patient with preganglionic lesion both pupils will dilate whereas in postganglionic lesions, the Horner's pupil will not. It is because, since the hydroxyamphetamine acts by releasing norepinephrine from the nerve endings at the myoneural junction, so in postganglionic lesions, the drug will have no effect.

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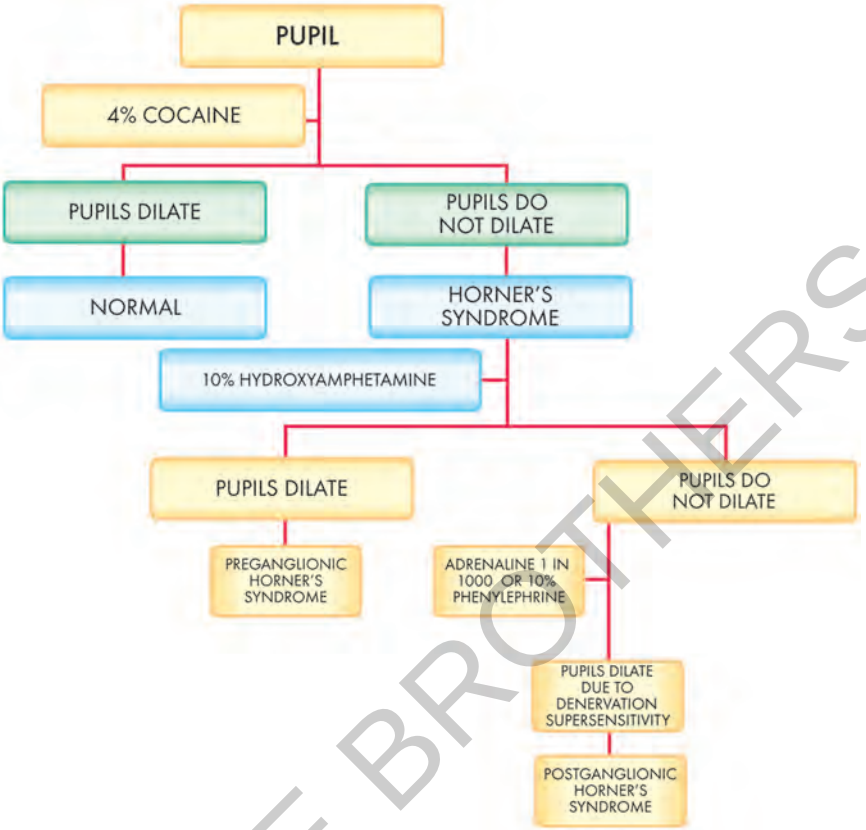


Figure 4.8 Pharmacological tests to localize Horner's syndrome

Adrenaline test or phenylephrine test: When either adrenaline 1 in 1000 or phenylephrine 10 percent is instilled in both eyes, the Horner's pupil due to postganglionic lesion dilates more than the normal pupil because of denervation hypersensitivity.

Reader's Syndrome

This applies to painful postganglionic Horner's syndrome. Cluster headaches are present. Raeder's paratrigeminal syndrome is a term that should probably be limited to the occasional middle fossa mass that produces trigeminal nerve involvement with pain and a postganglionic Horner's syndrome.

PHARMACOLOGY OF THE PUPIL

The iris has a sphincter controlled by the parasympathetic system (Fig. 4.9) and a dilator controlled by the sympathetic system (Fig. 4.10). In both, the systems acetylcholine is the neurotransmitter at the synapse between the preganglionic

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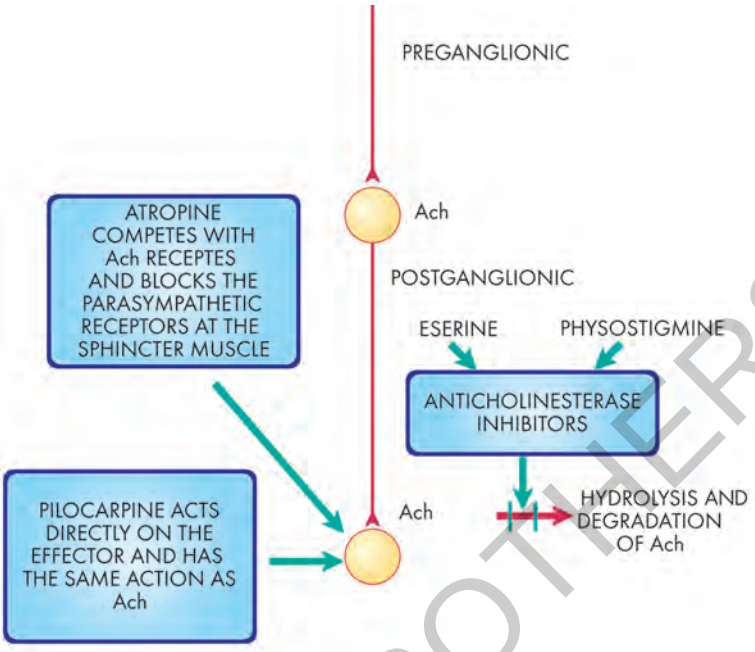


Figure 4.9 Parasympathetic system

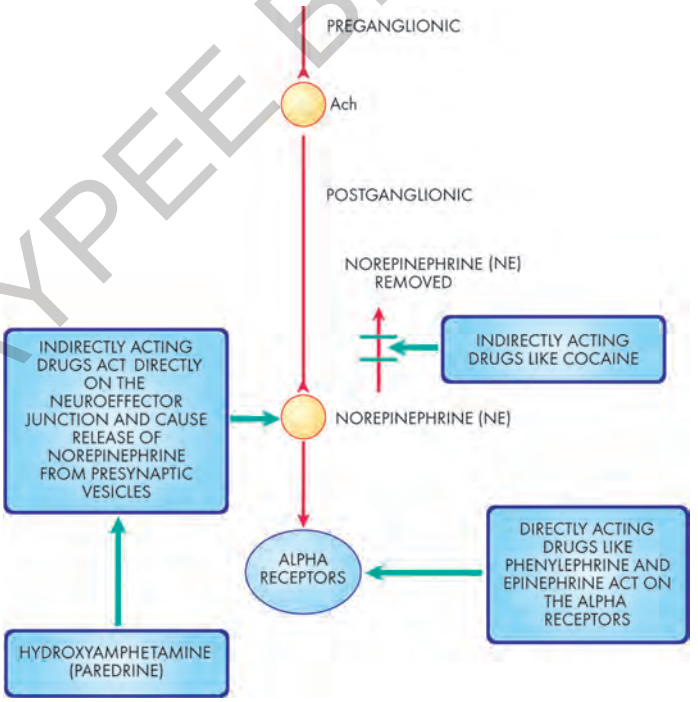


Figure 4.10 Sympathetic system

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and postganglionic neurons. In the parasympathetic system, acetylcholine is the neuroeffector at the sphincter. In the sympathetic system, the neuroeffector at the dilator is norepinephrine. There are alpha- and beta-sympathetic receptors in the ciliary body. The beta-receptors are inhibitory. Norepinephrine acts only upon alpha-receptors, which are the only receptors present in the dilator muscle of the iris. An iris deprived of postganglionic innervation develops supersensitivity within 24 to 48 hours, i.e. a physiologic response occurs when lesser amounts of cholinergic and sympathetic substances are present than those usually required to cause the same physiologic response.

Parasympathetic drugs can be of three types: (i) the directly acting drugs like pilocarpine which act directly on the effector and have the same action as the acetylcholine, (ii) indirect-acting cholinergic agents that interfere with the hydrolysis and degradation of acetylcholine, for example, the anticholinesterase inhibitors like eserine and physostigmine, and (iii) anticholinergic drugs that compete with acetylcholine for receptors at the effector site of the sphincter muscle, such as atropine. Atropine blocks the parasympathetic receptors at the sphincter muscle and prevents the muscle constriction caused by acetylcholine.

Sometimes, one has to find out whether a fixed dilated pupil (Fig. 4.11) is due to an interruption of parasympathetic innervation by an anatomic lesion or to pharmacological dilatation of the pupil. The examiner should instill 1 to 2 percent pilocarpine in the affected eye. If the patient has received an anticholinergic drug that dilated the pupil by blocking the receptor site, the pupil will not constrict. If the pupillary dilatation is due to interruption of the parasympathetic pathways proximal to the neuroeffector junction, the pilocarpine will act directly on the sphincter muscle and will cause pupillary constriction.

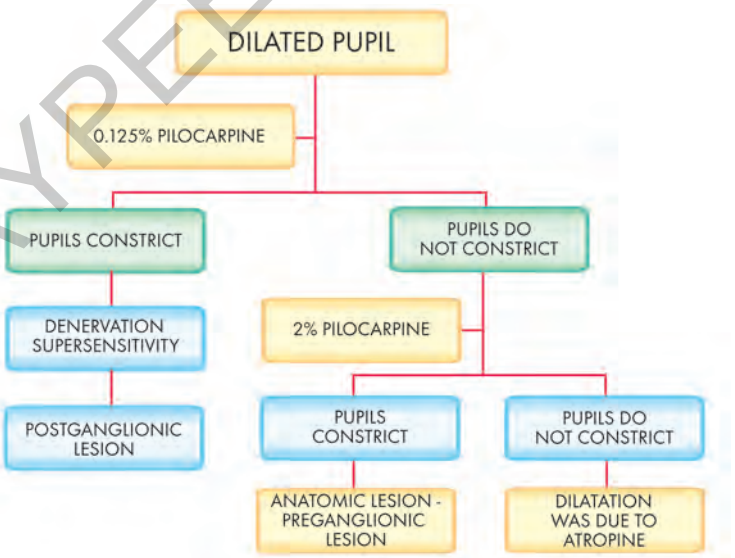


Figure 4.11 Dilated pupil

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Sympathetic acting drugs can be directly acting or indirectly acting drugs. The directly acting drugs like phenylephrine or epinephrine act directly on the alpha-receptors dilating the pupil. The indirectly acting drugs like cocaine and hydroxyamphetamine (Paredrine) act in a different way. Cocaine causes pupillary dilatation by blocking the normal re-uptake of constantly released norepinephrine. This leads to an accumulation of norepinephrine at the neuroeffector junction. Hydroxyamphetamine acts directly on the neuroeffector junction and causes release of norepinephrine from presynaptic vesicles. The presence of norepinephrine in presynaptic vesicles depends on the integrity of the third-order neuron. The presence of norepinephrine in these vesicles is not impaired by interruption of the preganglionic (second-order) neuron.

PUPIL ABNORMALITIES (FIGS 4.12 TO 4.15)

Hippus

Hippus is a visible rhythmic but irregular pupillary oscillation, deliberate in time and 2 mm or more in excursion. It has no localizing significance. It occurs in:

- Normal person
- Presence of total third nerve palsy
- Hemiplegia
- Multiple sclerosis
- Meningitis (acute)
- Cerebral syphilis
- Myasthenia gravis
- Epileptics.



Figure 4.12 Horner's syndrome—right eye. There is partial ptosis, miosis

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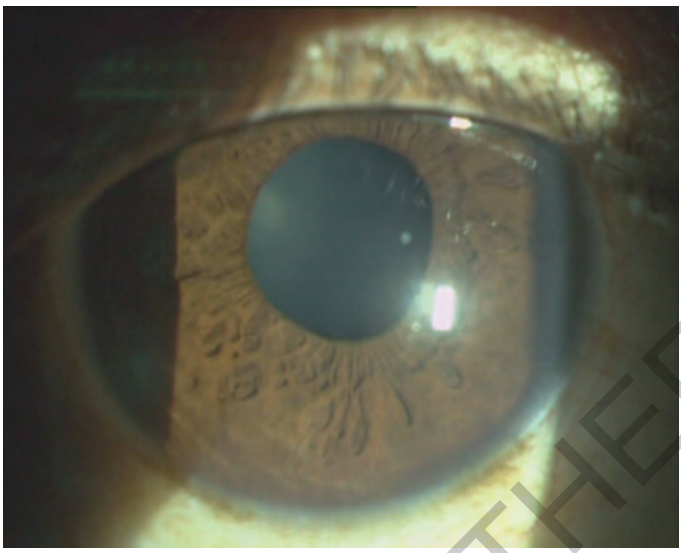


Figure 4.13 Adies pupil. Shows partial sphincteric action



Figure 4.14 Adies pupil before pilocarpine—left eye. Left eye pupil is dilated

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Paradoxical Pupillary Reaction

In this either: (i) the pupil dilates with near vision or constricts in distant vision, or (ii) the pupil dilates on exposure to light or constricts when the light is withdrawn. It occurs in:

- Syphilis
- Tumors of quadrigeminal region
- Sleeping individuals who have taken barbiturates
- Trauma.



Figure 4.15 Adies pupil after pilocarpine—left eye. Instilling 0.125 percent pilocarpine miosis occurs due to denervation supersensitivity

Irregularity of the Pupil

Irregularity of the pupil occurs in:

- Congenital coloboma of the iris
- Operations on the iris—like sector iridectomies
- Adherent leukoma as one part of the iris is pulled up to the corneal scar
- Peripheral anterior synechiae
- Iritis
- Glaucoma
- Tumors of the iris or ciliary body
- Argyll Robertson pupil
- Iridocorneal endothelial syndrome
- Optic atrophy.

Polycoria

In this, there are more than one pupils present. It occurs in

- Congenital
- Surgical—due to a surgical iridectomy
- Iridocorneal endothelial syndrome (essential iris atrophy)
- Iridoschisis.

REFERENCES

1. Agarwal S, Agarwal A, et al. Textbook of Ophthalmology, Vol. 4. Jaypee Brothers Medical Publishers, New Delhi, India. 2003.
2. Agarwal A. Handbook of Ophthalmology. Slack, USA, 2005.

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Amar Agarwal MS FRCS FRCOphth is the Chairman and Managing Director, Dr Agarwal's Group of Eye Hospitals and Eye Research Center, Chennai, Tamil Nadu, India. He is the Past President, International Society of Refractive Surgery (ISRS) and Secretary General, Indian Intraocular Implant and Refractive Society (IIRSI), India. He is the pioneer of Phakonit, which is Phaco with a Needle Incision Technology. This technique became popularized as bimanual phaco, microincision cataract surgery (MICS) or Microphaco. He is the first to remove cataracts through a 0.7 mm tip with the technique called Microphakonit. He has also discovered no anesthesia cataract surgery (NACS) and fallen vitreous (FAVIT) technique, a new technique to remove dropped nuclei. The air pump which was a simple idea of using an aquarium fish pump to increase the fluid into the eye in bimanual phaco, and co-axial phaco has helped prevent surge. He has discovered a new refractive error (Aberropia) and the first-to-do a combined surgery of microphakonit (700 micron cataract surgery) with a 25-gauge vitrectomy in the same patient. He is also the first surgeon to implant a new mirror telescopic IOL (LMI) for patients suffering from age-related macular degeneration. He has also been the first in the world to implant a Glued IOL, in which a PC-IOL is fixed in an eye without any capsules using fibrin glue. Dr Agarwal's Eye Hospital has done for the first time, an anterior segment transplantation in a 4-month-old child with anterior staphyloma. He has combined Glued IOL and IOL scaffold in cases of PC rupture where there is no iris or capsular support and termed the technique Glued IOL Scaffold. His hospital has done a glued endocapsular ring in cases of subluxated cataract.

He has started pre-Descemet's endothelial keratoplasty (PDEK), in which pre-Descemet's layer and the Descemet's membrane with endothelium are transplanted en bloc in patients with diseased endothelium. His hospital has also introduced contact lens-assisted collagen crosslinking (CACXL) for the first time, a new technique for crosslinking thin corneas, and has also worked on E-DMEK, in which an endoilluminator is used to assist in Descemet's membrane endothelial keratoplasty (DMEK) surgeries.

He has received many awards for his work done in ophthalmology, most significant being the Casebeer Award, Barraquer Award and the Kelman Award. He has performed more than 150 live surgeries at various conferences. His videos have won many awards at the film festivals of the ASCRS, AAO and ESCRS. He has also written more than 60 books, which have been published in various languages (English, Spanish and Polish). He also trains doctors from all over the world in his center on phaco, bimanual phaco, LASIK and vitreoretinal surgeries.

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