Eye Movement DISORDERS

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To my parents, Esther and Joseph,

and William, James, and Stephen

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Preface

Eye movement disorders are commonly encountered in clinical practice. They are often the initial manifestation of diseases affecting the central nervous system. Understanding eye movement disorders remains challenging, partly because it requires knowledge of the underlying anatomy and physiology. Although there are a number of excellent texts covering this subject, few present the information in a clear and concise manner with accompanying anatomic diagrams to elucidate the intricate relationships among clinical phenomena, basic neuroanatomy, and neurophysiology. With this book, I have attempted to fill this gap: text and illustrations are combined to provide a coherent and easy-to-assimilate description and explanation of different eye movement disorders.

The text is divided into four parts. The first part consists of chapters for each of the eye movement subsystems. Readers will find a description of thematic “concepts” on the left-hand pages, with accompanying figures, a synopsis of the pertinent points, and important clinical points (highlighted in green) on the right-hand pages. My aim is to present the material that appears on the left-hand pages as succinct, accessible information for easy review on the right-hand pages.

Parts II through IV describe different eye movement disorders in detail. Bulleted and numbered lists have been used to reduce the overall volume of the text without compromising the clarity of the information. Each chapter contains color-coded sections to allow ready review. The clinical features of different disorders are summarized in yellow boxes, etiology in green boxes, and differential diagnosis in orange boxes. Main eye movement abnormalities characteristic of different diseases are summarized in blue boxes, and the anatomic and physiological basis for the observed abnormal eye movements are explained separately, as footnotes. Throughout the text, readers will also find this icon ▸▸, which indicates that a video clip of the corresponding eye movement disorder is available in the book’s accompanying CD-ROM. In addition, the book is amply illustrated with schematic diagrams of relevant anatomy and brain pathways. Tables have also been used liberally to provide a readily accessible overview of information.

The book is comprehensive, though not exhaustive; I have aimed at a clear and understandable presentation of what I think are the most important aspects of eye movement disorders. I encourage readers to consult other excellent texts and the references provided for more detail on particular subjects of interest, particularly Leigh and Zee, *The Neurology of Eye Movements*, Fourth Edition, and Miller et al., *Walsh and Hoyt’s Clinical Neuro-Ophthalmology*, Sixth Edition. Readers who would like to view additional videos of different eye movement disorders can visit a number of web pages that are accessible to the public, such as the resourceful NOVEL website of the North American Neuro-Ophthalmology Society (http://library.med.utah.edu/NOVEL/), and the eTextbook of Eye Movements on the Canadian

Neuro-Ophthalmology Group website (www.neuroophthalmology.ca/textbook/NOeyemovt.html).

It is my hope that this book will serve as a resource for residents, fellows, and clinicians in different specialties: ophthalmology, neurology, neuro-ophthalmology, and neurosurgery. Neuro-otologists, orthoptists, medical students, as well as undergraduate and graduate students in behavioral neurosciences, should also find useful information here.

Acknowledgments

A number of individuals provided encouragement throughout this project and reviewed the manuscript critically. I would particularly like to thank Martin ten Hove, Barry Skarf, Lawrence Tychsen, Nancy Newman, Martin Steinbach, Carol Westall, Raymond Buncic, James Sharpe, Susan Culican, Daniel Weisbrod, Megumi Iizuka, Michael Richards, Peter Karagiannis, and Alan Blakeman. I would also like to express my gratitude to John Leigh and David Zee for writing an outstanding reference book on the neurology of eye movements; some information and many tables, particularly in Part III, included in this textbook are modified from their book, *The Neurology of Eye Movements*, Third Edition.

I am grateful to the past and present ophthalmology and neurology residents at the University of Toronto and Washington University in St. Louis for their feedback on my lectures notes, on which this book is based. I am also thankful to the PGY1 ophthalmology residents from across Canada who attended the Toronto Ophthalmology Residency Introductory Course for their support and suggestions. I am indebted to Mano Chandrakumar, who spent many hours helping me prepare the figures and the manuscript and who provided excellent technical support. Finally, I would like to thank the Canadian Institutes for Health Research, the E.A. Baker Foundation of the Canadian National Institute for the Blind, the Ontario Ministry of Research and Innovation, the Ophthalmology Practice Plan of the Toronto Western Hospital, the Department of Ophthalmology and Vision Sciences at the University of Toronto, and the Hospital for Sick Children in Toronto for their continued support of my work.

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Contents

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Abbreviations

AC	Anterior canal
A_g	Gravitational acceleration vector
A_i	Inertial (or linear translational) acceleration vector
ANA	Antinuclear antibody
ATD	Ascending tract of Deiters
BC	Brachium conjunctivum
BPPV	Benign paroxysmal positioning vertigo
CCN	Central caudal nucleus
CFEOM1	Congenital fibrosis of the extraocular muscles type 1
CFEOM2	Congenital fibrosis of the extraocular muscles type 2
CFEOM3	Congenital fibrosis of the extraocular muscles type 3
CHAMPS	Controlled High Risk Avonex Multiple Sclerosis Trial
CJD	Creutzfeldt-Jakob Disease
CMAPs	Compound muscle action potentials
cMRF	Central mesencephalic reticular formation
CNS	Central nervous system
CPEO	Chronic progressive external ophthalmoplegia
CSF	Cerebrospinal fluid
DLPC	Dorsolateral prefrontal cortex
DLPN	Dorsolateral pontine nuclei
DVD	Dissociated vertical deviation
EA-2	Episodic ataxia type 2
EBN	Excitatory burst neurons
EEG	Electroencephalography
EKG	Electrocardiogram
EMG	Electromyography
ERG	Electroretinogram
ESR	Erythrocyte sedimentation rate
EWN	Edinger-Westphal nucleus
FEF	Frontal eye field
FEFsac	Saccade subregion of the frontal eye field
FEFsem	Pursuit subregion of the frontal eye field
FL/VPF	Flocculus and ventral paraflocculus
FOR	Fastigial oculomotor region
FTN	Flocculus target neurons
GEN	Gaze-evoked nystagmus
GIA	Gravitoinertial acceleration vector
GPi	Globus pallidus internal segment
HAART	Highly active antiretroviral therapy
HC	Horizontal canal
HGPPS	Horizontal gaze palsy and progressive scoliosis
HT	Hypertropia
IBN	Inhibitory burst neurons
IML	Internal medullary lamina
INC	Interstitial nucleus of Cajal
INO	Internuclear ophthalmoplegia
IO	Inferior oblique
IR	Inferior rectus
IVIG	Intravenous immunoglobulin
LGN	Lateral geniculate nucleus
LIP	Lateral interparietal area
LP	Lumbar puncture
LR	Lateral rectus
LS	Lateral suprasylvian area
MBP	Myelin basic protein
mepps	Miniature endplate potentials
Med RF	Medullary reticular formation
MIF	Multiply innervated fibers
MLF	Medial longitudinal fasciculus
MOG	Myelin oligodendrocyte glycoprotein
MR	Medial rectus

MRF	Mesencephalic reticular formation
MSA-C	Multiple system atrophy dominated by cerebellar ataxia
MSA-P	Multiple system atrophy dominated by parkinsonism
MST	Medial superior temporal visual area
MT	Middle temporal visual area
MuSK	Muscle specific kinase
MVH-NPH	Medial vestibular nucleus - nucleus prepositus hypoglossi
MVN	Medial vestibular nucleus
NMDA	*N*-methyl-D-aspartate
NOT	Nucleus of the optic tract
NPC	Near point of convergence
nPC	Nucleus of the posterior commissure
NPH	Nucleus prepositus hypoglossi
NPH-MVN	Nucleus prepositus hypoglossi and medial vestibular nucleus complex
NRPC	Nucleus reticularis pontis caudalis
NRTP	Nucleus reticularis tegmenti pontis
OFR	Ocular following response
OKAN	Optokinetic after-nystagmus
OKN	Optokinetic nystagmus
ONTT	Optic Neuritis Treatment Trial
OPCA	Olivopontocerebellar atrophy
OTR	Ocular tilt reaction
PC	Posterior canal
PoC	Posterior commissure
PCR	Polymerase chain reaction
PD	Prism diopters
PEF	Parietal eye field
PGD	Nucleus paragigantocellularis dorsalis
PhyH	Phytanoyl-CoA hydroxyl-ase
PIVC	Parieto-insular-vestibular cortex
PLP	Proteolipid protein
PMT	Cell groups of the paramedian tracts
PPRF	Paramedian pontine reticular formation
PrP^{c}	Cellular proteinaceous infectious particle
PrP^{Sc}	Scrapie proteinaceous infectious particle
PSP	Progressive supranuclear palsy
PVN	Post-rotatory vestibular nystagmus
RAPD	Relative afferent pupillary defect
riMLF	Rostral interstitial nucleus of the medial longitudinal fasciculus
rip	Nucleus raphe interpositus
SC	Superior colliculus
SCA2	Spinocerebellar ataxia type 2
SCA6	Spinocerebellar ataxia type 6
SEF	Supplementary eye field
SIF	Singly innervated fibers
SNpc	Substantia nigra pars compacta
SNpr	Substantia nigra pars reticulata
SO	Superior oblique
SR	Superior rectus
SVN	Superior vestibular nucleus
SWJ	Square wave jerks
TMP-SMX	Trimethoprim-sulfamethoxazole
V_1	Ophthalmic division of the trigeminal nerve
V_2	Maxillary division of the trigeminal nerve
VN	Vestibular nystagmus
VOR	Vestibulo-ocular reflex
VPF	Ventral paraflocculus

Eye Movement Disorders

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Part I *The Six Eye Movement Systems*

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Chapter 1 *Eye Rotations, the Extraocular Muscles, and Strabismus Terminology*

To understand how eye muscles move the eyeball, it is necessary to understand the geometry of the eye and the functions of the muscles. The eyeball rotates about three axes: horizontal, vertical, and torsional. These axes intersect at the center of the eyeball. Eye rotations are achieved by coordinated contraction and relaxation of six extraocular muscles—four rectus and two oblique—attached to each eye. The action of the muscles on the globe is determined by the point of rotation of the globe, as well as the origin and insertion of each muscle. Recent evidence suggests that the muscles also exert their effects on the globe via the extraocular muscle pulleys.

Considering that we make at least 100,000 saccades alone each day, it is not surprising that many extraocular muscles are very resistant to fatigue. Extraocular muscles are also different from other skeletal muscles in many respects. For example, eye muscle fibers are richly innervated, and each motoneuron innervates only 10–20 muscle fibers, the smallest motor unit known in the body. Extraocular muscles also have more mitochondria and a higher metabolic rate than other skeletal muscles. Thus, extraocular muscles are one of the fastest contracting muscles. This property allows animals to shift gaze swiftly, so that they can avoid approaching predators or detect prey in the vicinity. The unique immunologic and physiologic properties of extraocular muscles may also explain why they are more susceptible to certain disease processes, such as Grave's disease and chronic progressive external ophthalmoplegia, but more resistant to others such as Duchenne's dystrophy, which mainly affects skeletal muscles in the rest of the body.

1.1 Three Axes of Eye Rotations

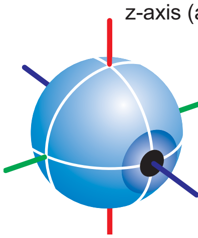
The eyeball rotates about three axes: x-axis (naso-occipital or roll axis), y-axis (earth-horizontal or pitch axis), and z-axis (earth-vertical or yaw axis).

Ductions refer to monocular movements of each eye. They include abduction, adduction, elevation (sursumduction), depression (deorsumduction), incycloduction or incyclotorsion, and excycloduction or excyclotorsion (see table on opposite page).

Versions refer to binocular conjugate movements of both eyes, such that the visual axes of the eyes move in the same direction. They include dextroversion, levoversion, elevation (sursumversion), depression (deorsumversion), dextrocycloversion, and levocycloversion (see table).

Vergences refer to binocular disjunctive movements, such that the visual axes of the eyes move in opposite directions. They include convergence, divergence, incyclovergence, and excyclovergence (see table).

Eye rotations are achieved by coordinated contraction of six extraocular muscles in each eye: the medial rectus, lateral rectus, superior rectus, inferior rectus, superior oblique, and inferior oblique. The action of the muscles on the globe is determined by the point of rotation of the globe, as well as by the origin and insertion of each muscle. The tendons of the rectus muscles pass through sleevelike pulleys located several millimeters posterior to the equator of the globe and approximately 10 mm posterior to the insertion sites of the muscles. These pulleys, consisting of fibrous tissue and smooth muscle, limit side-slip movement of the rectus muscles during eye rotations and act as the functional origins of the rectus muscles.



Term	Definition
Ductions	Ductions refer to **monocular** movements of each eye ▪ **Abduction** occurs about the z-axis and is away from the median plane ▪ **Adduction** occurs about the z-axis and is toward the median plane ▪ **Elevation** occurs about the y-axis and is an upward rotation of the eye ▪ **Depression** occurs about the y-axis and is a downward rotation of the eye ▪ **Incycloduction** (incyclotorsion) occurs about the x-axis so that the upper pole of the eye rotates toward the median plane ▪ **Excycloduction** (excyclotorsion) occurs about the x-axis so that the upper pole of the eye rotates away from the median plane
Versions	Versions refer to binocular **conjugate** movements of both eyes, such that the visual axes of the eyes move in the same direction ▪ **Dextroversion**: both eyes rotating about their z-axes to the right ▪ **Levoversion**: both eyes rotating about their z-axes to the left ▪ **Elevation**: both eyes rotating about their y-axes to look upward ▪ **Depression**: both eyes rotating about their y-axes to look downward ▪ **Dextrocycloversion**: both eyes rotating about their x-axes so that the upper pole of both eyes rotates to the subject's right ▪ **Levocycloversion**: both eyes rotating about their x-axes so that the upper pole of both eyes rotates to the subject's left
Vergences	Vergences refer to binocular **disjunctive** movements, such that the visual axes of the eyes move in opposite directions ▪ **Convergence**: both eyes rotating about their z-axes toward the median plane ▪ **Divergence**: both eyes rotating about their z-axes away from the median plane ▪ **Incyclovergence**: rotation of both eyes about their x-axes so that the upper pole of both eyes rotates toward the median plane ▪ **Excyclovergence**: rotation of both eyes about their x-axes so that the upper pole of both eyes rotates away from the median plane

1.2 Actions of the Extraocular Muscles

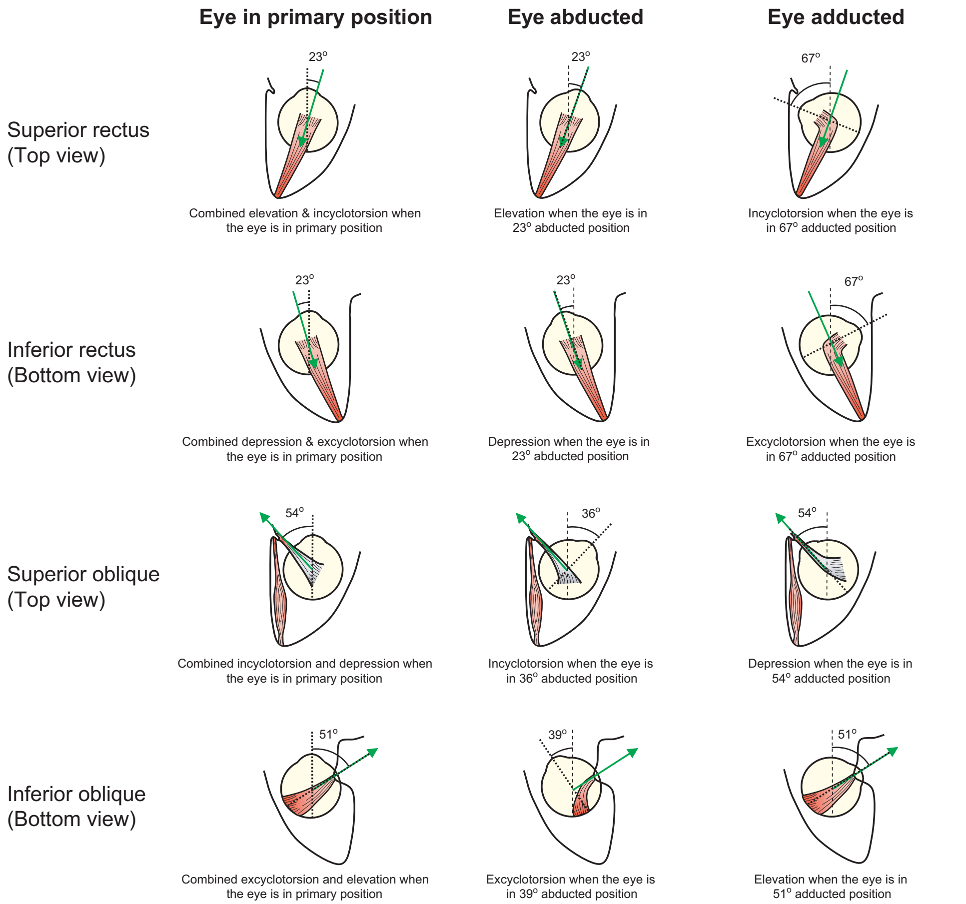
The primary position of the eye is defined clinically as the position in which the eye is directed straight ahead with the body and head erect. The primary action of a muscle is its major rotational effect on the eye while the eye is in primary position. The secondary and tertiary actions of a muscle are additional rotational effects on the eye while it is in primary position.

The four rectus muscles arise from the annulus of Zinn at the apex of the orbit. The **medial rectus** inserts onto the medial side of the globe at approximately 5.3 (±0.7) mm from the corneoscleral limbus, whereas the **lateral rectus** inserts onto the lateral side of the globe at approximately 6.9 (±0.7) mm from the limbus. Because the origins and insertions of the horizontal rectus muscles are symmetric and lie in the horizontal meridian of the globe, their functions are relatively simple and are antagonistic; contraction of the medial rectus adducts the globe, whereas contraction of the lateral rectus abducts the globe.

The superior and inferior recti also originate from the annulus of Zinn. The **superior rectus** inserts onto the globe superiorly at approximately 7.9 (±0.6) mm from the limbus, and the **inferior rectus** inserts inferiorly at approximately 6.8 (±0.8) mm from the limbus. In addition, their insertions onto the globe subtend a 23° angle with the visual axis when the eye is in primary position, straddling the vertical meridian of the globe. Thus, in addition to its primary action of elevation, the superior rectus has a secondary action of incyclotorsion and a tertiary action of adduction. The primary action of the inferior rectus is depression, its secondary action is excyclotorsion, and its tertiary action is adduction. The relative importance of the primary and secondary actions depends on the direction of the visual axis. When the eye is abducted 23°, the superior rectus acts solely as an elevator, and the inferior rectus acts solely as a depressor. When the eye is adducted 67°, the superior rectus acts solely to incyclotort the globe, and the inferior rectus acts solely to excyclotort the globe.

The **superior oblique** also arises from the annulus of Zinn; however, its functional origin is the trochlea in the superomedial orbit. The superior oblique is tendinous after it passes through the trochlea. This tendon then assumes a posterolateral direction and inserts onto the superior posterotemporal quadrant of the globe behind the center of rotation. This vector plane subtends a 54° angle with the visual axis when the eye is in primary position. Thus, in addition to its primary action of incyclotorsion, the superior oblique also has a secondary action of depression and tertiary action of abduction. When the eye is adducted 54°, the superior oblique acts solely to depress the globe, and when the eye is abducted 36°, it acts solely to incyclotort the globe.

The **inferior oblique** arises from the anterior medial orbital floor, and thus it is the only extraocular muscle that does not arise from the annulus of Zinn. It inserts onto the inferior posterotemporal quadrant of the globe behind the center of rotation and subtends a 51° angle with the visual axis when the eye is in primary position. Thus, in addition to its primary action of excyclotorsion, the inferior oblique has a secondary action of elevation and a tertiary action of abduction. When the eye is adducted 51°, the inferior oblique acts solely to elevate the globe, and when the eye is abducted 39°, it acts solely to excyclotort the globe.



Extraocular Muscles	Primary Action	Secondary Action	Tertiary Action
Lateral rectus	Abduction	None	None
Medial rectus	Adduction	None	None
Superior rectus	Elevation	Incyclotorsion	Adduction
Inferior rectus	Depression	Excyclotorsion	Adduction
Superior Oblique	Incyclotorsion	Depression	Abduction
Inferior oblique	Excyclotorsion	Elevation	Abduction

1.3 Laws of Ocular Motor Control and the Six Cardinal Positions of Gaze

Agonist muscle moves the eye toward the desired direction, whereas antagonist muscle moves the eye away from the desired direction. **Sherrington's law of reciprocal innervation** states that, whenever an agonist muscle (e.g., the medial rectus of the right eye during adduction) receives an excitatory signal to contract, an equivalent inhibitory signal is sent to the antagonist muscle (e.g., the right lateral rectus) of the same eye. This reciprocal innervation is mainly due to central connections in the brainstem.

A yoked muscle pair consists of one muscle from each eye and moves both eyes toward the same direction. For example, the right lateral rectus and the left medial rectus contract simultaneously when looking to the right. **Hering's law of equal innervation** (or law of motor correspondence) states that, during conjugate eye movements, the yoked muscle pair receives equal innervation so that the eyes move together. Vertically acting muscles are also conceptualized as being arranged as yoked pairs (e.g., the right superior rectus and left inferior oblique form a pair, and the right inferior rectus and the left superior oblique form another pair). However, the way in which extraocular muscles interact is very complex, and all muscles probably contribute, even during a simple horizontal movement.

During clinical examination, the **primary position** refers to the position when the eyes look straight ahead. Secondary positions are right gaze, left gaze, straight up, and straight down. Tertiary positions are up and right, down and right, up and left, and down and left.

The **six cardinal positions** include right gaze, left gaze, and the four tertiary positions. These eye positions provide the most information about the horizontal function of the horizontal rectus muscles (lateral and medial rectus) and the vertical function of the cyclovertical muscles (superior rectus, inferior rectus, superior oblique, and inferior oblique). For example, on right and up gaze, the prime elevators are the superior rectus in the right eye and the inferior oblique in the left eye. In this gaze position, the right superior rectus is the prime elevator when the right eye is abducted (by the action of the lateral rectus) because it inserts at a 23° angle to the visual axis. Similarly, the left inferior oblique is the prime elevator when the left eye is adducted (by the action of the medial rectus) because it inserts at a 51° angle to the visual axis.

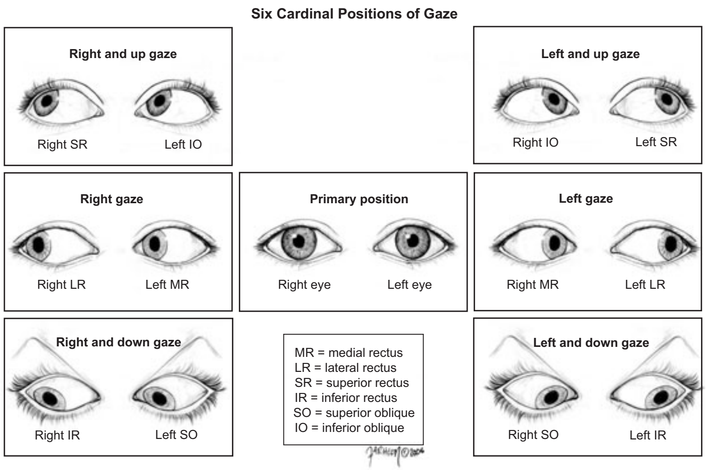
It is important to emphasize that the cardinal positions of gaze do not correspond to the primary, secondary, or tertiary actions of the muscles. For example, when the right eye looks right and up, the right superior rectus is not responsible for *both* elevation *and* abduction; in fact, the tertiary action of the superior rectus is adduction, not abduction. In other words, when the right eye looks right and up, elevation comes mainly from contraction of the superior rectus, whereas abduction comes mainly from contraction of the lateral rectus.

Sherrington's law of reciprocal innervation

Whenever an agonist muscle (e.g., the medial rectus of the right eye during adduction) receives an excitatory signal to contract, an equivalent inhibitory signal is sent to the antagonist muscle (e.g., the right lateral rectus) of the same eye.

Hering's law of equal innervation

During conjugate eye movements, the yoked muscle pair receives equal innervation so that the eyes move together.



Clinical Points

The six cardinal positions of gaze are the eye positions that provide the most information about:

- The horizontal function of the horizontal muscles (lateral and medial rectus)
- The vertical function of the cyclovertical muscles (superior rectus, inferior rectus, superior oblique, and inferior oblique).

For example, on right and upgaze:

- The right superior rectus (SR) is most responsible for elevation when the right eye is *abducted* because the SR inserts at a 23° angle to the visual axis when the eye is in primary position. Note that the tertiary action of the superior rectus is *adduction*, not abduction. Thus, the cardinal position does not correspond to the action of the muscle; rather, it corresponds to the position of the eye that gives the most information about the vertical function of the cyclovertical muscle.
- The left inferior oblique (IO) is most responsible for elevation when the left eye is *adducted* because the IO inserts at a 51° angle to the visual axis when the eye is in primary position. Note that the tertiary action of the inferior oblique is *abduction*, not adduction.

1.4 Structure and Function of Extraocular Muscle Fibers

The rectus and oblique muscles consist of two distinct layers: an outer **orbital layer** adjacent to the periorbita and orbital bone, and an inner **global layer** adjacent to the eye and the optic nerve. Whereas the global layer extends the full muscle length, inserting via a well-defined tendon, the orbital layer ends before the muscle becomes tendinous. Each layer contains fibers more suited for sustained contraction or for brief, rapid contraction. Six types of fibers have been identified in the extraocular muscles.

In the orbital layer, about 80% of the fibers are **singly innervated fibers** (SIF). Not only do these fibers exhibit the fast type of myofibrillar ATPase and high oxidative activity, but they also appear to be capable of anaerobic activity. They have twitch capacity and are the most fatigue-resistant fibers. They are the only fiber type that shows long-term effects after injection of botulinum toxin. The remaining 20% of orbital fibers are **multiply innervated fibers** (MIF). They have twitch capacity near the center of the fiber and non-twitch activity proximal and distal to the endplate band.

In the global layer, about 33% of fibers are **red SIFs**, which are fast twitch and highly fatigue resistant. Another 32% are **white (pale) SIFs** with fast-twitch properties but low fatigue resistance. **Intermediate SIFs** constitute about another 25% of fibers. They have fast-twitch properties and an intermediate level of fatigue resistance. The remaining 10% are **MIFs**, with synaptic endplate along their entire length, as well as at the myotendinous junction, where there are palisade organ proprioceptors. These fibers show tonic properties, with slow, graded, nonpropagated responses to neural or pharmacological activation.

The **levator palpebrae** muscle contains the three singly innervated muscle types found in the global layer of the extraocular muscles and a true slow-twitch fiber type. The MIF type and the fatigue-resistant SIF type seen in the orbital layer of the extraocular muscles are absent in the levator.

The functional arrangement of muscle fiber types is related to the threshold at which motor units are recruited. During saccades and quick phases of nystagmus, all fiber types are recruited synchronously. In contrast, during slow eye movements and fixation (gaze holding), there is a differential recruitment of fiber types that is dependent on eye position. Orbital SIFs and global red SIFs are recruited first, when eye position is still in the direction opposite to the muscle action. Multiply innervated fiber types are recruited next, probably near straight-ahead position, where their fine increments of force would be of value for fixation. The increasingly faster but fatigable fibers are recruited last, at eye position well into the direction of muscle action.

The **palisade tendon organs** are the primary proprioceptors in human extraocular muscles. They are found in the distal myotendinous junctions of global MIFs. Afferents from these proprioceptors project, via the ophthalmic branch of the trigeminal nerve and the Gasserian ganglion, to the spinal trigeminal nucleus. They may also project centrally via the ocular motor nerves. From the trigeminal nucleus, proprioceptive information is sent to structures involved in ocular motor control, including the superior colliculus, vestibular nuclei, nucleus prepositus hypoglossi, cerebellum, and frontal eye fields. Proprioceptive information is also sent to structures involved in visual processing, including the lateral geniculate body, pulvinar, and visual cortex.

	Orbital layer		Global layer			
	Orbital SIF	Orbital MIF	Global Red SIF	Global White SIF	Global Intermediate SIF	Global MIF
% of layer	80	20	33	32	25	10
Contraction mode	Twitch	Mixed	Twitch	Twitch	Twitch	Non-twitch
Contraction speed	Fast	Fast/slow	Fast	Fast	Fast	Slow
Fatigue resistance	High	Variable	High	Low	Intermediate	High
Recruitment order *Slow eye movements and fixation*	1st	3rd	2nd	6th	5th	4th
Saccades and quick phases of nystagmus	All fiber types recruited simultaneously					

SIF, singly innervated fibers; MIF, multiply innervated fibers. (Modified from Porter et al. Extraocular muscles: Basic and clinical aspects of structure and function. Surv Ophthelmol. 1995; 39: 451–84.)

The **palisade tendon organs**

- Primary proprioceptors in human extraocular muscles
- Found in the distal myotendinous junctions of global multiply innervated fibers
- Project to the spinal trigeminal nucleus via the ophthalmic branch of the trigeminal nerve and the Gasserian ganglion, or via the ocular motor nerves
- From the spinal trigeminal nucleus, proprioceptive information is sent to

1. Structures involved in ocular motor control (e.g., superior colliculus, vestibular nuclei, nucleus prepositus hypoglossi, cerebellum, frontal eye fields)
2. Structures involved in visual processing (e.g., lateral geniculate body, pulvinar, visual cortex).

1.5 Strabismus Terminology

The line connecting the object of fixation to the fovea is the **visual axis**. **Strabismus** is defined as a misalignment of the visual axes between the two eyes.

Orthophoria is the ideal condition of eye alignment. In reality, it is seldom encountered because the majority of people have a latent misalignment. By definition, orthophoria indicates that the oculomotor apparatus is in perfect equilibrium so that both eyes remain aligned in all positions of gaze and at all distances of fixation during viewing with one eye (monocular viewing). **Orthotropia** refers to perfect alignment of the eyes during viewing with both eyes (binocular viewing).

Heterodeviation refers to ocular alignment that differs from orthophoria. It includes both **heterophoria** and **heterotropia**. Heterophoria is a latent deviation controlled by binocular fixation, such that, during viewing with both eyes, the eyes remain aligned. In contrast, heterotropia is a deviation present during viewing with both eyes (i.e., manifest deviation).

There are a variety of heterophoric and heterotropic deviations. If the visual axes converge, the condition is called **esophoria** (for latent deviation) or **esotropia** (for manifest deviation). If the visual axes diverge, the condition is known as **exophoria** or **exotropia**. **Uncrossed diplopia** is double vision caused by esotropia. The false image is displaced on the same side as the deviated eye. **Crossed diplopia** is double vision caused by exotropia. The false image is displaced to the side opposite the deviated eye.

Hyperphoria (for latent deviation) or **hypertropia** (for manifest deviation) occurs if the visual axis of the nonfixating eye is higher than that of the fixating eye. For example, a right hyperphoria or hypertropia is a deviation in which the visual axis of the nonfixating right eye is higher than that of the left. **Cyclodeviation** is a torsional misalignment of the eyes, causing a cyclodisparity. **Incyclodeviation** refers to a relative incyclotorsion of the eyes (decreased separation of upper poles of eyes), whereas **excyclodeviation** refers to a relative excyclotorsion of the eyes (increased separation of upper poles of eyes).

Strabismus may be comitant or incomitant. In **concomitant** or comitant strabismus, the magnitude of deviation is the same in all directions of gaze and does not depend on the eye used for fixation. In **incomitant** or noncomitant strabismus, the deviation varies in different directions of gaze. Most incomitant strabismus is caused by a paralytic or a mechanical restrictive process. The deviation is largest when the eyes turn in the direction of the paralytic or underacting muscle. The deviation in incomitant strabismus also varies with the eye used for fixation. When the normal eye is fixating, the amount of misalignment is called **primary deviation**. When the paretic eye is fixating, the amount of misalignment is called **secondary deviation**. Secondary deviation is larger than primary deviation in incomitant strabismus because an increase in innervation is needed for a paretic eye to fixate a target. By Hering's law, the contralateral yoked muscle also receives more innervation, resulting in a larger deviation.

Weakness of a muscle can be classified as a paralysis or paresis. If the action of a muscle is completely abolished, the condition is a paralysis or palsy; if the action of a muscle is weakened but not abolished, it is called a paresis. The terms palsy and paresis are often used interchangeably in clinical settings and in neurologic practice. In this book, the term palsy is used to denote a partial or a complete impairment of muscle action.

Term	Definition
Visual axis	The line connecting the object of fixation to the fovea
Strabismus	A misalignment or deviation of the visual axes
Orthophoria	Alignment of the visual axes while viewing with one eye
Orthotropia	Alignment of the visual axes while viewing with both eyes
Heterophoria	A latent misalignment of the visual axes while viewing with one eye
Heterotropia	A manifest misalignment of the visual axes while viewing with both eyes
Esophoria/esotropia	Convergence of visual axes (i.e., crossed eyes) during viewing with one eye (esophoria) or during viewing with both eyes (esotropia)
Exophoria/exotropia	Divergence of visual axes (i.e., walled eyes) during viewing with one eye (exophoria) or during viewing with both eyes (exotropia)
Hyperphoria/hypertropia	Vertical misalignment of the visual axes with the nonfixating eye higher than the fixating eye during viewing with one eye (hyperphoria) or during viewing with both eyes (hypertropia)
Hypophoria/hypotropia	Vertical misalignment of the visual axes with the nonfixating eye lower than the fixating eye during viewing with one eye (hypophoria) or during viewing with both eyes (hypotropia)
Cyclodeviation	Torsional misalignment of the eyes, causing a cyclodisparity
Incyclodeviation	Relative incyclotorsion of the eyes (decreased separation of upper poles of eyes)
Excyclodeviation	Relative excyclotorsion of the eyes (increased separation of upper poles of eyes)
Uncrossed diplopia	Double vision caused by esotropia; the false image is displaced on the same side as the deviated eye
Crossed diplopia	Double vision caused by exotropia; the false image is displaced to the side opposite to the deviated eye
Concomitant deviation	Misalignment of the visual axes that does not change with gaze direction during fixation with either eye
Incomitant deviation	Misalignment of the visual axes that changes with gaze direction and depends on which eye is fixating; causes include mechanical restriction or muscle palsy
Primary deviation	The deviation of the paretic eye while the normal eye is fixating
Secondary deviation	The deviation of the normal eye while the paretic eye is fixating; secondary deviation is larger than primary deviation in incomitant strabismus

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Chapter 2 Introduction to the Six Eye Movement Systems and the Visual Fixation System

One main reason that we make eye movements is to solve a problem of information overload. A large field of vision allows an animal to survey the environment for food and to avoid predators, thus increasing its survival rate. Similarly, a high visual acuity also increases survival rates by allowing an animal to aim at a target more accurately, leading to higher killing rates and more food. However, there are simply not enough neurons in the brain to support a visual system that has high resolution over the entire field of vision. Faced with the competing evolutionary demands for high visual acuity and a large field of vision, an effective strategy is needed so that the brain will not be overwhelmed by a large amount of visual input. Some animals, such as rabbits, give up high resolution in favor of a larger field of vision (rabbits can see nearly 360°), whereas others, such as hawks, restrict their field of vision in return for a high visual acuity (hawks have vision as good as 20/2, about 10 times better than humans). In humans, rather than using one strategy over the other, the retina develops a very high spatial resolution in the center (i.e., the fovea), and a much lower resolution in the periphery. Although this "foveal compromise" strategy solves the problem of information overload, one result is that unless the image of an object of interest happens to fall on the fovea, the image is relegated to the low-resolution retinal periphery.

The evolution of a mechanism to move the eyes is therefore necessary to complement this foveal compromise strategy by ensuring that an object of interest is maintained or brought to the fovea. To maintain the image of an object on the fovea, the vestibulo-ocular (VOR) and optokinetic systems generate eye movements to compensate for head motions. Likewise, the saccadic, smooth pursuit, and vergence systems generate eye movements to bring the image of an object of interest on the fovea. These different eye movements have different characteristics and involve different parts of the brain. In this chapter, the fixation system is discussed; the VOR and optokinetic systems, saccades, smooth pursuit, and vergence systems are discussed in subsequent chapters.