

THE NEUROLOGY OF EYE MOVEMENTS

FOURTH EDITION

R. John Leigh, M.D., F.R.C.P.

*Blair-Daroff Professor of Neurology,
Professor of Neurosciences, Otolaryngology,
and Biomedical Engineering
Case Western Reserve University
Department of Veterans Affairs Medical Center
and University Hospitals
Cleveland, Ohio*

David S. Zee, M.D.

*Professor of Neurology, Ophthalmology,
Otolaryngology and Head and
Neck Surgery, and Neuroscience
The Johns Hopkins University School of Medicine
Baltimore, Maryland*

OXFORD
UNIVERSITY PRESS
2006

Contents

Part 1 The Properties and Neural Substrate of Eye Movements

1. A SURVEY OF EYE MOVEMENTS: CHARACTERISTICS AND TELEOLOGY 3

- Why Study Eye Movements? 3
- Visual Requirements of Eye Movements 5
- Functional Classes of Eye Movements 5
- Orbital Mechanics: Phasic and Tonic Innervation 6
- Vestibular and Optokinetic Systems 8
- Saccadic System 10
- Smooth Pursuit and Visual Fixation 11
- Combined Movements of the Eyes and Head 12
- Vergence Eye Movements 12
- Three-Dimensional Aspects of Eye Movements 13
- Adaptive Control of Eye Movements 13
- Voluntary Control of Eye Movements 14
- Eye Movements and Spatial Localization 14
- The Scientific Method Applied to the Study of Eye Movements 15
- Summary 16

2. THE VESTIBULAR-OPTOKINETIC SYSTEM 20

- Function of the Vestibular-Optokinetic System 22
- Anatomy and Physiology of the Peripheral Vestibular System 25
- Brainstem Elaboration of the Vestibulo-ocular Reflex 31
- Neural Substrate for Optokinetic Responses 40
- Quantitative Aspects of the Vestibular-Optokinetic System 40
- Adaptive Properties of the Vestibulo-ocular Reflex 53
- Vestibulocerebellar Influences on the Vestibulo-ocular Reflex 59
- Vestibular Sensation and the Role of the Cerebral Hemispheres in the Vestibulo-ocular Reflex 62
- Clinical Examination of Vestibular and Optokinetic Function 64
- Laboratory Evaluation of Vestibular and Optokinetic Function 71
- Pathophysiology of Disorders of the Vestibular System 76
- Summary 81

3. THE SACCADIC SYSTEM 108

- The Purpose of Saccades 108
- Behavior of the Saccadic System 109
- Neurophysiology of Saccadic Eye Movements 124
- Models for Saccade Generation 149
- Adaptive Control of Saccadic Accuracy 153
- Saccades and Movements of the Eyelids 156
- Examination of Saccades 158
- Pathophysiology of Saccadic Abnormalities 160
- Summary 165

4. SMOOTH PURSUIT AND VISUAL FIXATION 188

- The Purpose of Smooth Pursuit 188
- Visual Fixation 189
- Stimulus for Smooth Pursuit 193
- Quantitative Aspects of Smooth Pursuit 197
- Adaptive Properties of Smooth Pursuit 201
- Neural Substrate for Smooth Pursuit 202
- Models of Smooth Pursuit 214
- Clinical Examination of Fixation and Smooth Pursuit 217
- Laboratory Evaluation of Fixation and Smooth Pursuit 218
- Abnormalities of Visual Fixation and Smooth Pursuit 219
- Summary 226

5. GAZE HOLDING AND THE NEURAL INTEGRATOR 241

- Neural Coding of the Ocular Motor Signal 242
- Quantitative Aspects of Neural Integration 245
- Neural Substrate for Gaze Holding 246
- Clinical Evaluation of Gaze Holding 253
- Abnormalities of the Neural Integrator: Gaze-Evoked Nystagmus 254
- Summary 256

6. SYNTHESIS OF THE COMMAND FOR CONJUGATE EYE MOVEMENTS 261

- Hypothetical Pathways for Gaze Control 262
- Brainstem Connections for Horizontal Conjugate Movements 262
- Brainstem Connections for Vertical and Torsional Movements 268
- Cerebellar Influences on Gaze 274
- The Cerebral Hemispheres and Voluntary Control of Eye Movements 280
- Descending, Parallel Pathways That Control Voluntary Gaze 295
- Summary 299

7. EYE-HEAD MOVEMENTS 315

- Ocular Motor and Cephalomotor Systems 315
- Stabilization of the Head 315
- Voluntary Control of Eye-Head Movements 317
- Examination of Eye-Head Movements 327
- Laboratory Evaluation of Eye-Head Movements 328
- Disorders of Eye-Head Movement 329
- Summary 335

8. VERGENCE EYE MOVEMENTS 343

- The Purpose of Vergence Eye Movements 343
- Stimuli to Vergence Movements 344
- Dynamic Properties of Vergence Eye Movements 348
- Neural Substrate of Vergence Movements 355
- Conceptual Models of Supranuclear Control of Vergence 360
- Adaptive Mechanisms to Maintain Ocular Alignment 361
- Examination of Vergence Movements 365
- Laboratory Testing of Vergence Eye Movements 367
- Abnormalities of Vergence 367
- Summary 372

Part 2 The Diagnosis of Disorders of Eye Movements**9. DIAGNOSIS OF PERIPHERAL OCULAR MOTOR PALSIES AND STRABISMUS 385**

- A Pathophysiological Approach to Diplopia 386
- Anatomy of the Orbital Fascia and the Extraocular Muscles 387
- Structure and Function of Extraocular Muscle 392
- Anatomy of Ocular Motor Nerves and Their Nuclei 396
- Physiologic Basis for Conjugate Movements: Yoke Muscle Pairs 401
- Clinical Testing in Diplopia 402
- Pathophysiology of Some Commonly Encountered Signs in Strabismus 409
- Clinical Features and Diagnosis of Concomitant Strabismus 414
- Clinical Features of Ocular Nerve Palsies 416
- Disorders of the Neuromuscular Junction 438
- Chronic Progressive External Ophthalmoplegia and Restrictive Ophthalmopathies 444

10. DIAGNOSIS OF NYSTAGMUS AND SACCADIC INTRUSION 475

- The Nature and Visual Consequences of Abnormal Eye Movements That Prevent Steady Fixation 476

Clinical and Laboratory Methods for Evaluating Nystagmus and Saccadic Intrusions	476
A Pathophysiological Approach to the Diagnosis of Nystagmus	479
Nystagmus Due to Vestibular Imbalance	480
Periodic Alternating Nystagmus	493
Seesaw and Hemi-Seesaw Nystagmus	496
Nystagmus Occurring When the Eyes are in Eccentric Gaze	498
Nystagmus Occurring in Association with Disease of the Visual System	502
Acquired Pendular Nystagmus and its Relationship to Disease of the Visual Pathways	505
Convergent-Divergent Forms of Nystagmus	511
Congenital Forms of Nystagmus	512
Lid Nystagmus	521
Saccadic Intrusions	521
Treatments for Nystagmus and Saccadic Intrusions	528

11. DIAGNOSIS AND MANAGEMENT OF VESTIBULAR DISORDERS 559

Vertigo and Dizziness	559
Oscillopsia	578
Skew Deviation and the Ocular Tilt Reaction (OTR)	581

12. DIAGNOSIS OF CENTRAL DISORDERS OF OCULAR MOTILITY 598

Using Eye Movements for Topological Diagnosis	599
Ocular Motor Syndromes Caused by Lesions in the Medulla	599
Ocular Motor Syndromes Caused by Disease of the Cerebellum	605
Ocular Motor Syndromes Caused by Disease of the Pons	616
Ocular Motor Syndromes Caused by Lesions of the Midbrain	630
Ocular Motor Syndromes Caused by Lesions in the Superior Colliculus	645
Ocular Motor Syndromes Caused by Lesions in the Diencephalon	645
Ocular Motor Abnormalities and Disease of the Basal Ganglia	648
Ocular Motor Syndromes Caused by Lesions in the Cerebral Hemispheres	654
Abnormalities of Eye Movements in Patients with Dementia	670
Eye Movement Disorders in Psychiatric Illnesses	671
Eye Movements in Stupor and Coma	673
Ocular Motor Dysfunction and Multiple Sclerosis (MS)	678
Ocular Motor Manifestations of Metabolic and Deficiency Disorders	680
Effects of Drugs on Eye Movements	683

APPENDIX A 719**APPENDIX B 722****INDEX 727****INSTRUCTIONS FOR USING THE DVD 763**