

Contents

Introduction to the BCSC	xv
Introduction to Section 5	xvii
Objectives	1
1 Neuro-Ophthalmic Anatomy	3
Highlights	3
Introduction	3
Bony Anatomy	4
Skull Base	4
The Orbit	5
Vascular Anatomy	10
Arterial System	10
Anterior Circulation	12
Posterior Circulation	18
Venous System	20
Afferent Visual Pathways	24
Retina	24
Optic Nerve	25
Optic Chiasm	27
Optic Tract	27
Cortex	29
Efferent Visual System	32
Cortical Input	33
Subcortical Structures	34
Brainstem	35
Ocular Motor Cranial Nerves	37
Extraocular Muscles	45
Sensory and Facial Motor Anatomy	45
Fifth Cranial Nerve	45
Seventh Cranial Nerve	49
Eyelids	51
Ocular Autonomic Pathways	52
Sympathetic Pathways	52
Parasympathetic Pathways	55
2 Imaging in Neuro-Ophthalmology	57
Highlights	57
Introduction	57
Computed Tomography	59

Magnetic Resonance Imaging	59
Crucial Considerations in Neuroimaging	66
When to Order	69
What to Order	69
How to Order	70
How to Interpret	71
Vascular and Other Imaging Modalities	72
Catheter Angiography	72
Magnetic Resonance Angiography and Magnetic Resonance Venography	73
Computed Tomography Angiography and Computed Tomography Venography	73
Metabolic and Functional Imaging Modalities	75
Ultrasonography	75
Optical Coherence Tomography	76
Glossary of Select Neuroimaging Terminology	84
3 The Patient With Decreased Vision: Evaluation	87
Highlights	87
History	87
Unilateral Versus Bilateral Involvement	87
Time Course of Vision Loss	88
Associated Signs and Symptoms	88
Cognitive Status	88
Examination	89
Visual Acuity Testing	89
Color Vision Testing	90
Pupillary Testing	91
Fundus Examination	94
Visual Field Evaluation	96
Examination: Adjunct Testing	102
Contrast Sensitivity Testing	102
Photostress Recovery Testing	104
Potential Acuity Meter Testing	104
Fluorescein and Indocyanine Green Angiography	104
Fundus Autofluorescence	105
Optical Coherence Tomography	107
Ultrasonography	115
Electrophysiologic Testing	117
4 The Patient With Decreased Vision Due to Retinal, Optic Nerve, and Chiasmal Diseases	125
Highlights	125
Introduction	125
Ocular Media Abnormalities	126
Retinopathy	126
AIBSE Syndrome, AZOOR, and MEWDS	127
Cone Dystrophy	128
Autoimmune Retinopathies and Paraneoplastic Retinopathies	129

Optic Neuropathy	131
Visual Field Patterns in Optic Neuropathy	131
Causes of Optic Neuropathy	132
Chiasmal Lesions	180
Visual Field Loss Patterns	181
Etiology of Chiasmal Disorders	181
5 The Patient With Visual Dysfunction Due to Retrochiasmal Disease	187
Highlights	187
Introduction	187
Retrochiasmal Lesions	187
Optic Tract	188
Lateral Geniculate Nucleus	189
Optic Radiations of the Temporal Lobe	190
Optic Radiations of the Parietal Lobe	191
Optic Radiations of the Occipital Lobe	191
Cortical Visual Impairment	194
Vision Rehabilitation	195
6 The Patient With Transient Visual Loss	197
Highlights	197
Clinical Characteristics	197
Examination	199
Monocular Transient Visual Loss	200
Ocular Causes	200
Orbital Causes	200
Vascular Causes	202
Binocular Transient Visual Loss	211
Migraine	211
Occipital Mass Lesions	211
Occipital Ischemia	211
Occipital Seizures	212
7 The Patient With Illusions, Hallucinations, and Disorders of Higher Cortical Function	213
Highlights	213
Introduction	213
Visual Distortion and Illusions	215
Ocular Origin	215
Optic Nerve Origin	216
Cortical Origin	216
Hallucinations	216
Ocular Origin	216
Optic Nerve Origin	217
Cortical Origin	217
Treatment of Illusions and Hallucinations	220

Disorders of Higher Cortical Function	220
Disorders of Recognition	221
Disorders of Visual–Spatial Relationships	223
Disorders of Awareness of Vision or Visual Deficit	224

8 The Patient With Abnormal Ocular Motility or Diplopia	227
Highlights	227
Introduction	227
History	228
Physical Examination	229
Monocular Diplopia	231
Differentiating Paretic From Restrictive Causes of Diplopia	231
Comitant and Incomitant Deviations	232
Localization of Potential Lesions	232
Supranuclear Causes of Abnormal Ocular Motility	233
Nuclear Causes of Diplopia	234
Internuclear Causes of Diplopia	235
Internuclear Ophthalmoplegia	235
One-and-a-Half Syndrome	236
Ocular Motor Cranial Nerve Palsies	237
Intra-axial Ocular Motor Cranial Nerve Palsies	238
Third Cranial Nerve Palsy	239
Fourth Cranial Nerve Palsy	244
Sixth Cranial Nerve Palsy	246
Recurrent Painful Ophthalmoplegic Neuropathy	248
Paresis of More Than One Cranial Nerve	248
Cavernous Sinus and Orbital Apex Involvement	248
Myopathic, Restrictive, Orbital, Neuromuscular, and Other Causes of Diplopia	252
Thyroid Eye Disease	252
Orbital Myositis	252
Posttraumatic Restriction	252
Postsurgical Restriction	253
Chronic Progressive External Ophthalmoplegia	254
Neoplastic Involvement	254
Brown Syndrome	254
Heavy Eye Syndrome	254
Sagging Eye Syndrome	254
Ocular Neuromyotonia	255
Neuromuscular Junction Causes of Diplopia	255
Refractive Procedure–Induced Diplopia	255
Foveal Displacement Syndrome	255

9 The Patient With Supranuclear Disorders of Ocular Motility	257
Highlights	257
Fundamental Principles of Ocular Motor Control	257
Supranuclear Ocular Motor Systems: Function, Anatomy, Clinical Testing, and Disorders of Eye Movements	259
Ocular Fixation System	259
Vestibular-Ocular System (Vestibular-Ocular Reflex)	260
Optokinetic System (Optokinetic Nystagmus)	267
Saccadic System	268
Smooth-Pursuit System	276
Vergence System	279
Gaze Preference, Gaze Palsy, and Tonic Deviations	282
10 The Patient With Nystagmus or Spontaneous Eye Movement Disorders	287
Highlights	287
Introduction and Terminology	287
Assessment of Eye Movement Abnormalities	288
Early-Onset (Childhood) Nystagmus	289
Infantile Nystagmus Syndrome (Congenital Nystagmus)	289
Fusional Maldevelopment Nystagmus Syndrome (Latent Nystagmus)	291
Heimann-Bielschowsky Phenomenon	293
Spasmus Nutans Syndrome	294
Gaze-Evoked Nystagmus	295
Rebound Nystagmus	296
Vestibular Nystagmus	296
Peripheral Vestibular Nystagmus	296
Central Vestibular Nystagmus	299
HINTS Test	302
Acquired Pendular Nystagmus	303
Oculopalatal Myoclonus or Tremor	303
See-Saw Nystagmus	304
Dissociated Nystagmus With Internuclear Ophthalmoplegia	304
Saccadic Intrusions	305
Saccadic Intrusions With Normal Intersaccadic Intervals	305
Saccadic Intrusions Without Normal Intersaccadic Intervals	306
Voluntary Flutter (Voluntary “Nystagmus”)	307
Additional Eye Movement Disorders	307
Convergence-Retraction Nystagmus	307
Superior Oblique Myokymia	308
Oculomasticatory Myorhythmia	309
11 The Patient With Pupillary Abnormalities	311
Highlights	311
Introduction	311

History	311
Pupillary Examination	312
Factors Affecting Pupil Size	313
Pupil Irregularity	313
Anisocoria	314
Normal Pupillary Response to Light	314
Anisocoria Equal in Dim and Bright Light	314
Anisocoria Greater in Dim Light	316
Anisocoria Greater in Bright Light	322
Disorders of Pupillary Reactivity: Light–Near Dissociation	325
Afferent Visual Pathway	325
Midbrain	325
Aberrant Regeneration of Cranial Nerve III	326
Other Pupillary Disorders	326
Benign Episodic Unilateral Mydriasis	326
12 The Patient With Eyelid or Facial Abnormalities	327
Highlights	327
Introduction	327
Examination Techniques	328
Ptosis	331
Congenital Ptosis	331
Acquired Ptosis	332
Pseudoptosis	333
Apraxia of Eyelid Opening	334
Eyelid Retraction	334
Abnormalities of Facial Movement	335
Seventh Cranial Nerve Disorders	337
Disorders of Underactivity of the Seventh Cranial Nerve	337
Disorders of Overactivity of the Seventh Cranial Nerve	342
13 The Patient With Head, Ocular, or Facial Pain	347
Highlights	347
Head Pain	347
Headache Associated With Systemic Conditions	348
Migraine and Tension-type Headache	350
Trigeminal Autonomic Cephalgias and Hemicrania Continua	357
Idiopathic Stabbing Headache	358
Inherited Encephalopathies Resembling Migraine	358
Ocular and Orbital Pain	359
Troclear Headache and Trochleitis	360
Photophobia	361
Facial Pain	361
Herpes Zoster Ophthalmicus	361
Internal Carotid Artery Dissection	362
Giant Cell Arteritis	362
Neoplastic Processes	362

Trigeminal Neuralgia	362
Occipital Neuralgia	363
14 The Patient With Functional Neurological Symptom (Conversion) and Related Disorders	365
Highlights	365
Introduction	366
Clinical Profile	368
Examination Techniques.	369
Afferent Visual Pathway	369
Ocular Motility and Alignment	377
Pupils and Accommodation	378
Eyelid Position and Function	379
Management of the Patient With Functional Neurological Symptom and Related Disorders	379
Functional Neurological Symptom (Conversion) Disorder	379
Somatic Symptom Disorder.	380
15 The Patient With a Systemic Disease	381
Highlights	381
Introduction	381
Immunologic Disorders	381
Giant Cell Arteritis	382
Multiple Sclerosis	384
Neuromyelitis Optica Spectrum Disorder and Myelin Oligodendrocyte Glycoprotein Immunoglobulin G-Associated Disorder.	389
Myasthenia Gravis	394
Thyroid Eye Disease	397
Sarcoidosis	397
Inherited Disorders With Neuro-Ophthalmic Signs	398
Myopathies.	398
Neurocutaneous Syndromes	400
Selected Neuro-Ophthalmic Disorders Associated With Pregnancy	403
Posterior Reversible Encephalopathy Syndrome	405
Lymphocytic Hypophysitis	405
Cerebrovascular Disorders	406
Transient Visual Loss	406
Vertebobasilar System Disease	406
Cerebral Aneurysms.	409
Arterial Dissection	412
Arteriovenous Malformations.	413
Cerebral Venous Thrombosis	415
Reversible Cerebral Vasoconstriction Syndrome.	417
Neuro-Ophthalmic Manifestations of Infectious Diseases	419
Human Immunodeficiency Virus Infection	419
Herpesvirus	421

<i>Mycobacterium</i>	421
Syphilis	422
Progressive Multifocal Leukoencephalopathy	422
Toxoplasmosis	422
Lyme Disease	423
Fungal Infections	424
Prion Diseases	428
Radiation Therapy	428
Additional Materials and Resources	431
Requesting Continuing Medical Education Credit	433
Study Questions	435
Answers	445
Index	455