

Contents

Introduction to the BCSC	xv
Objectives	1
1 Basic Concepts in Immunology:	
Effector Cells and the Innate Immune Response	3
Highlights	3
Definitions	3
Cellular Components of the Immune System	4
Neutrophils	5
Eosinophils	5
Basophils and Mast Cells	5
Monocytes and Macrophages	6
Dendritic Cells and Langerhans Cells	7
Lymphocytes	7
Overview of the Innate Immune System	8
Immunity Versus Inflammation	8
Innate Immunity: Triggers and Mechanisms	10
Bacteria-Derived Molecules That Trigger Innate Immune Responses	10
Other Triggers or Modulators of Innate Immune Responses	12
Innate Mechanisms for the Recruitment and Activation of Neutrophils	13
Innate Mechanisms for the Recruitment and Activation of Macrophages	15
Phagocyte Killing Mechanisms	18
Mediator Systems That Amplify Immune Responses	19
Plasma-Derived Enzyme Systems	20
Histamine	21
Lipid Mediators	21
Cytokines	23
Neutrophils and Their Products	27
2 Immunization and Adaptive Immunity:	
The Immune Response Arc and Immune Effectors	31
Highlights	31
Definitions	31
Phases of the Immune Response Arc	32
Afferent Phase	32
Processing Phase	34
Effector Phase	38

The Immune Response Arc and Primary or Secondary Immune Response	39
Differences Between Primary and Secondary Responses	39
Effector Responses and Mechanisms of Adaptive Immunity	40
Antibody-Mediated Effector Responses	41
Lymphocyte-Mediated Effector Responses	44
Combined Antibody and Cellular Effector Mechanisms	48
3 Ocular Immune Responses	51
Highlights	51
Regional Immunity and Immunologic Microenvironments	51
Immune Responses of the Conjunctiva	53
Features of the Immunologic Microenvironment	53
Immunoregulatory Systems	53
Immune Responses of the Cornea	54
Features of the Immunologic Microenvironment	54
Immunoregulatory Systems	55
Immune Responses of the Anterior Chamber, Anterior Uvea, and Vitreous	56
Features of the Immunologic Microenvironment	56
Immunoregulatory Systems	56
Immune Responses of the Retina, Retinal Pigment Epithelium, Choriocapillaris, and Choroid	57
Features of the Immunologic Microenvironment	57
Immunoregulatory Systems	58
4 Special Topics in Ocular Immunology	61
Highlights	61
Animal Models of Human Uveitis	61
Human Leukocyte Antigen Associations and Disease	62
Major Histocompatibility Complex and Human Leukocyte Antigen Molecules	62
Allelic Variation	63
Disease Associations	64
5 Diagnostic Considerations in Uveitis	67
Highlights	67
Overview	67
Epidemiology	68
Classification of Uveitis	69
Anatomical Classification	70
Classification by Clinical Features	72
Symptoms of Uveitis	73
Signs of Uveitis	73
Anterior Segment	74
Intermediate Segment	78
Posterior Segment	80
Review of the Patient's Health and Other Associated Factors	80

Differential Diagnosis of Uveitis	81
Ancillary Testing	86
Ophthalmic Imaging and Functional Tests	89
Ocular Fluid and Tissue Sampling	93
Cytology and Pathology	94
6 Therapy for Uveitis	97
Highlights	97
Introduction	97
Medical Management of Uveitis	97
Corticosteroids	99
Systemic Immunomodulatory Therapy	110
Other Therapeutic Agents	122
Surgical Management of Uveitis	122
7 Scleritis	123
Highlights	123
Introduction	123
Classification of Scleritis	123
Pathophysiology	124
Epidemiology	124
Clinical Presentation	125
Diffuse Anterior Scleritis	126
Nodular Anterior Scleritis	126
Necrotizing Scleritis	127
Posterior Scleritis	130
Infectious Scleritis	130
Diagnosis	131
Treatment	133
Systemic Treatment	134
Surgical Treatment	135
Prognosis	136
8 Noninfectious Anterior and Intermediate Uveitis	137
Highlights	137
Anterior Uveitis	137
Acute Anterior Uveitis	138
Chronic Anterior Uveitis	151
Intermediate Uveitis	157
Pars Planitis	157
Multiple Sclerosis	162
9 Posterior Uveitis: The White Dot Syndromes	165
Highlights	165
Definitions	165
Overview of the White Dot Syndromes	166
Birdshot Chorioretinopathy	167
Acute Posterior Multifocal Placoid Pigment Epitheliopathy	175

Serpiginous Choroiditis	178
Ampiginous Choroiditis, or Relentless Placoid Chorioretinitis	180
Multifocal Choroiditis With Panuveitis, Punctate Inner Choroiditis, and Subretinal Fibrosis and Uveitis Syndrome	182
Multiple Evanescent White Dot Syndrome	188
Acute Retinal Pigment Epitheliitis	192
Acute Zonal Occult Outer Retinopathy.	193
Acute Idiopathic Maculopathy	194
Autoimmune Retinopathy	196
Manifestations	196
Diagnosis	197
Treatment	198
10 Posterior Uveitis and Panuveitis With Possible Systemic Manifestations	199
Highlights	199
Introduction	199
Posterior Uveitis With Possible Systemic Manifestations	200
Systemic Lupus Erythematosus	200
Polyarteritis Nodosa	203
Granulomatosis With Polyangiitis and Microscopic Polyangiitis	204
Susac Syndrome	206
Panuveitis With Possible Systemic Manifestations	208
Sarcoidosis	208
Sympathetic Ophthalmia	214
Vogt-Koyanagi-Harada Syndrome.	217
Behçet Disease	223
11 Infectious Uveitis: Bacterial Causes	229
Highlights	229
Syphilis	229
Congenital Syphilis	230
Acquired Syphilis	230
Diagnosis	235
Treatment	236
Tuberculosis	237
Ocular Involvement	238
Diagnosis	241
Treatment	242
Lyme Disease	243
Clinical Features	244
Ocular Involvement	245
Diagnosis	245
Treatment	246
Leptospirosis	246
Ocular Involvement	247
Diagnosis	247
Treatment	247

Nocardiosis	248
Ocular Involvement	248
Diagnosis	248
Treatment	249
Bartonellosis	249
Ocular Involvement	249
Diagnosis	251
Treatment	251
Whipple Disease	252
Ocular Involvement	252
Diagnosis	253
Treatment	253
12 Infectious Uveitis: Nonbacterial Causes	255
Highlights	255
Viral Uveitis	255
Herpesviridae Family	255
Rubella	267
Lymphocytic Choriomeningitis Virus	269
Mumps	270
Measles (Rubeola)	270
West Nile Virus	272
Rift Valley Fever	273
Human T-cell Lymphotropic Virus Type 1	274
Dengue Fever	274
Chikungunya Fever	276
Zika Virus	277
Ebola Virus	279
SARS-CoV-2 and COVID-19	279
Other Viral Diseases	280
Fungal Uveitis	280
Ocular Histoplasmosis Syndrome	281
Coccidioidomycosis	283
Protozoal Uveitis	284
Toxoplasmosis	284
Helminthic Uveitis	292
Toxocariasis	292
Cysticercosis	295
Diffuse Unilateral Subacute Neuroretinitis	296
Onchocerciasis	300
13 Ocular Involvement in HIV Infection and AIDS	303
Highlights	303
Acquired Immunodeficiency Syndrome	303
Ophthalmic Manifestations	303
Cytomegalovirus Retinitis	304
Necrotizing Herpetic Retinitis	307
<i>Toxoplasma</i> Retinochoroiditis	307
Ocular Syphilis	309

Multifocal Choroiditis and Systemic Dissemination	309
External Eye Manifestations	311
14 Endophthalmitis	315
Highlights	315
Definitions	315
Chronic Postoperative Endophthalmitis	316
Clinical Findings	316
Diagnosis	317
Treatment	318
Endogenous Endophthalmitis	318
Endogenous Bacterial Endophthalmitis	318
Endogenous Fungal Endophthalmitis	320
15 Masquerade Syndromes	327
Highlights	327
Introduction	327
Neoplastic Masquerade Syndromes	327
Vitreoretinal Lymphoma	328
Uveal Lymphoma	332
Ocular Manifestations of Systemic Lymphoma	333
Ocular Manifestations of Leukemia	333
Nonlymphoid Tumors	333
Metastatic Tumors	335
Bilateral Diffuse Uveal Melanocytic Proliferation	335
Nonneoplastic Masquerade Syndromes	335
Retinitis Pigmentosa	335
Ocular Ischemic Syndrome	336
Chronic Peripheral Rhegmatogenous Retinal Detachment	336
Intraocular Foreign Bodies	336
Pigment Dispersion Syndrome	337
Infectious Uveitis	337
16 Complications of Uveitis	339
Highlights	339
Calcific Band Keratopathy	339
Cataracts	339
Management	340
Complications	342
Glaucoma	342
Uveitic Ocular Hypertension	343
Uveitic Glaucoma	343
Corticosteroid-Induced Ocular Hypertension and Glaucoma	344
Management	345
Hypotony	346
Uveitic Macular Edema	347
Treatment	347
Epiretinal Membrane and Macular Hole	349

Vitreous Opacification and Vitritis	349
Rhegmatogenous Retinal Detachment	350
Choroidal and Retinal Neovascularization	350
Vision Rehabilitation	351
Appendix A: Diagnostic Survey for Uveitis	353
Appendix B: Antimicrobial Agents for Intraocular Injection	357
Additional Materials and Resources	359
Requesting Continuing Medical Education Credit	361
Study Questions	363
Answers	371
Index	379