

Contents

1	Fundamentals	1	3.4.3	Muscle Tone and Strength	39
1.1	Microscopic Anatomy of the Nervous System	3	3.4.4	Reflexes	40
1.1.1	Neurons	3	3.4.5	Sensation	46
1.1.2	Neuroglia	3	3.5	Trunk	48
1.1.3	Myelin Sheaths	4	3.5.1	Back and Spine	48
1.1.4	Synapses	4	3.5.2	Reflexes	49
1.2	Elements of Neurophysiology	5	3.5.3	Sensation	49
1.2.1	Ion Channels	5	3.6	Lower Limbs	49
1.2.2	Resting Potential	5	3.6.1	General Aspects	49
1.2.3	Action Potential	5	3.6.2	Coordination and Strength	50
1.2.4	Impulse Conduction	5	3.6.3	Reflexes	50
1.3	Elements of Neurogenetics	5	3.6.4	Sensation	52
1.3.1	General Genetics	6	3.7	Autonomic Nervous System	52
1.3.2	Neurogenetics	7	3.8	Neurologically Relevant Aspects of the General Physical Examination	52
1.3.3	Genetic Counseling	8	3.9	neuropsychological and Psychiatric Examination	53
2	The Clinical Interview in Neurology	11	3.9.1	Psychopathologic Findings	53
2.1	General Principles of History-Taking	13	3.9.2	neuropsychological Examination	53
2.1.1	General Prerequisites for Good History-Taking	13	4	Ancillary Tests in Neurology	61
2.1.2	General Principles of the Clinical Interview	13	4.1	Fundamentals	63
2.1.3	Your Demeanor toward the Patient	13	4.2	Imaging Studies	63
2.1.4	The History and Physical Examination	13	4.2.1	Conventional Skeletal Radiographs	63
2.2	Special Aspects of History-Taking	14	4.2.2	Computed Tomography	64
2.2.1	The Present Illness	14	4.2.3	Magnetic Resonance Imaging	65
2.2.2	Past Medical History, Family History, and Social History	15	4.2.4	Angiography with Radiologic Contrast Media (Digital Subtraction Angiography)	68
3	The Neurologic Examination	17	4.2.5	Myelography and Radiculography	73
3.1	Basic Principles of the Neurologic Examination	19	4.2.6	Diagnostic Techniques of Nuclear Medicine	73
3.2	Stance and Gait	19	4.3	Electrophysiologic Studies	74
3.2.1	General Remarks	19	4.3.1	Fundamentals	74
3.2.2	Special Stance and Gait Tests	22	4.3.2	Electroencephalography	75
3.3	Head and Cranial Nerves	23	4.3.3	Evoked Potentials	78
3.3.1	Head and Cervical Spine	23	4.3.4	Electromyography	79
3.3.2	Cranial Nerves	25	4.3.5	Electroneurography	81
3.4	Upper Limbs	38	4.3.6	Other Electrophysiologic Studies	82
3.4.1	General Aspects	38	4.4	Ultrasonography (Neurosonography)	83
3.4.2	Motor Function and Coordination	38	4.5	Other Ancillary Studies	84
			4.5.1	Cerebrospinal Fluid Studies	84
			4.5.2	Tissue Biopsies	86
			4.5.3	Perimetry	88

5	Topical Diagnosis and Differential Diagnosis of Neurologic Syndromes	89	6.2.3	Assessment of Severity; Imaging Studies	121
5.1	Fundamentals	91	6.2.4	Pathophysiology and Clinical Features	122
5.2	Muscle Weakness and Other Motor Disturbances	91	6.2.5	Traumatic Hematomas	125
5.2.1	Anatomic Substrate of Motor Function	91	6.2.6	The Treatment of Traumatic Brain Injury	126
5.2.2	Motor Regulatory Systems	91	6.2.7	Complications	127
5.3	Sensory Disturbances	96	6.2.8	Prognosis	129
5.3.1	Anatomic Substrate of Sensation	96	6.3	Intracranial Pressure	129
5.3.2	The Peripheral Part of the Somatosensory System	97	6.3.1	Definition, Etiology, and Pathogenesis	129
5.3.3	The Central Part of the Somatosensory System	97	6.3.2	Clinical Features and Diagnostic Evaluation	130
5.4	Disturbances of Consciousness	99	6.3.3	Complication: Herniation	131
5.4.1	Somnolence, Stupor, and Coma: Severity and Causes	99	6.3.4	Treatment	131
5.4.2	The Differential Diagnosis of Coma	102	6.4	Brain Tumors	132
5.5	Dysfunction of Specific Areas of the Brain	102	6.4.1	Overview	132
5.5.1	Syndromes of the Individual Lobes of the Cerebral Hemispheres	102	6.4.2	Astrocytoma and Glioblastoma	135
5.5.2	Syndromes of the Extrapyramidal Motor System	104	6.4.3	Ependymoma	136
5.5.3	Thalamic Syndromes	105	6.4.4	Medulloblastoma and Primitive Neuroectodermal Tumors	136
5.5.4	The Limbic System	105	6.4.5	Oligodendroglioma	137
5.5.5	Brainstem Syndromes	106	6.4.6	Meningioma	137
5.5.6	Cerebellar Syndromes	107	6.4.7	Lymphoma	138
6	Diseases of the Brain and Meninges	111	6.4.8	Pituitary Tumors	138
6.1	Congenital and Perinatally Acquired Diseases of the Brain	113	6.4.9	Malformations and Hamartomatous Tumors	139
6.1.1	Fundamentals	113	6.4.10	Neurinomas	139
6.1.2	Cerebral Movement Disorders	113	6.4.11	Brain Metastases	140
6.1.3	Hydrocephalus	113	6.5	Cerebral Ischemia and Ischemic Stroke	140
6.1.4	Microcephaly	115	6.5.1	Overview	140
6.1.5	Dysraphic Malformations	115	6.5.2	Anatomy and Pathophysiology	141
6.1.6	Heterotopia	115	6.5.3	The Classification of Cerebral Ischemia by Severity	144
6.1.7	Ulegyria	115	6.5.4	Etiology, Risk Factors, and Primary Prophylaxis	145
6.1.8	Phakomatoses	116	6.5.5	The Dynamic Time Course of Cerebral Ischemia	148
6.1.9	Brain Infections Acquired in Utero	116	6.5.6	Infarct Types	148
6.1.10	Other Embryopathies	117	6.5.7	Clinical Stroke Syndromes	150
6.1.11	Malformations of the Skull and Craniocervical Junction	118	6.5.8	Diagnostic Evaluation	152
6.1.12	Mental Disorders	118	6.5.9	Treatment of Ischemic Stroke	154
6.2	Traumatic Brain Injury	119	6.5.10	Special Types of Cerebral Ischemia	156
6.2.1	Overview	119	6.6	Nontraumatic Intracranial Hemorrhage	156
6.2.2	Clinical History and Neurologic Examination	121	6.6.1	Intracerebral Hemorrhage	157
			6.6.2	Subarachnoid Hemorrhage	160
			6.7	Infectious Diseases of the Brain and Meninges	163
			6.7.1	Overview	163
			6.7.2	Acute Bacterial Meningitis	165
			6.7.3	Acute Viral Meningitis: Aseptic or Lymphocytic Meningitis	167
			6.7.4	Chronic Meningitis	167

6.7.5	Bacterial (Meningo)encephalitis: Spirochetal Infections	169	6.12.6	Vascular Dementia: SAE-Associated Dementia and Multi-Infarct Dementia	210
6.7.6	Viral Encephalitis	171	6.12.7	Dementia due to Malresorptive Hydrocephalus	211
6.7.7	Fungal Encephalitis	173			
6.7.8	Parasitic and Protozoal Encephalitis	173	7	Diseases of the Spinal Cord	213
6.7.9	Encephalitis in Prion Diseases: Creutzfeldt–Jakob Disease	175	7.1	Overview	215
6.7.10	Slow Virus Diseases	176	7.1.1	Anatomy	215
6.7.11	Intracranial Abscesses	177	7.1.2	The Main Spinal Cord Syndromes and Their Anatomic Localization	215
6.8	Metabolic Disorders and Systemic Diseases Affecting the Nervous System	178	7.1.3	Further Diagnostic Evaluation of Spinal Cord Lesions	219
6.8.1	Congenital Metabolic Disorders	178	7.2	Spinal Cord Trauma	220
6.8.2	Intoxications and Alcohol-Induced Disturbances of the Nervous System	182	7.3	Slowly Progressive Spinal Cord Compression	222
6.8.3	Endocrine Diseases	182	7.3.1	Spinal Cord Tumors	222
6.8.4	Gastrointestinal Diseases	183	7.3.2	Myelopathy due to Cervical Spondylosis	224
6.8.5	Hematologic Diseases	185	7.3.3	Syringomyelia and Syringobulbia	224
6.8.6	Collagen Diseases and Immune Diseases	185	7.4	Spinal Cord Ischemia and Hemorrhage	225
6.8.7	Paraneoplastic Syndromes	186	7.4.1	Blood Supply of the Spinal Cord	226
6.8.8	Limbic Encephalitis	186	7.4.2	Arterial Hypoperfusion	226
6.8.9	Renal Failure and Electrolyte Disturbances	186	7.4.3	Impaired Venous Drainage	227
6.9	Parkinson Disease and Other Hypertonic–Hypokinetic Syndromes	188	7.4.4	Hemorrhage in or adjacent to the Spinal Cord	227
6.9.1	Overview	188	7.5	Infectious and Inflammatory Diseases of the Spinal Cord	228
6.9.2	Parkinson Disease (Idiopathic Parkinson Syndrome)	188	7.5.1	Myelitis	228
6.9.3	Symptomatic Parkinsonian Syndromes	195	7.5.2	Spinal Abscesses	229
6.9.4	Progressive Supranuclear Palsy	195	7.6	Diseases Mainly Affecting the Long Tracts of the Spinal Cord	229
6.9.5	Multisystem Atrophy	195	7.6.1	Overview	229
6.9.6	Corticobasal Degeneration	196	7.6.2	Friedreich Ataxia	230
6.9.7	Lewy Body Dementia	197	7.6.3	Familial Spastic Spinal Paralysis	230
6.10	Chorea, Athetosis, Ballism, Dystonia: Hyperkinetic Syndromes	197	7.6.4	Funicular Myelosis	230
6.10.1	Huntington Chorea	197	7.7	Diseases of the Anterior Horns	231
6.10.2	Chorea Minor (Sydenham Chorea)	197	7.7.1	Overview	231
6.10.3	Athetosis	199	7.7.2	Spinal Muscular Atrophies	232
6.10.4	Ballism	199	7.7.3	Amyotrophic Lateral Sclerosis	232
6.10.5	Dystonic Syndromes	199			
6.10.6	Essential Tremor and Other Types of Tremor	200	8	Multiple Sclerosis and Other Autoimmune Diseases of the Central Nervous System	235
6.11	Cerebellar Diseases and Other Conditions Causing Ataxia	201	8.1	Fundamentals	237
6.11.1	Overview	201	8.2	Multiple Sclerosis	237
6.11.2	Selected Types of Ataxia	202	8.3	Other Autoimmune Diseases of the CNS	245
6.12	Dementia	203	8.3.1	Neuromyelitis Optica	245
6.12.1	Overview: The Dementia Syndrome	205	8.3.2	Acute Disseminated Encephalomyelitis	246
6.12.2	Alzheimer Disease (Senile Dementia of Alzheimer Type)	206			
6.12.3	Treatment and Prognosis	209			
6.12.4	Dementia with Lewy Bodies	209			
6.12.5	Frontotemporal Dementia (Pick Disease)	209			

8.3.3	Behçet Disease	246	10.3.4	Other Causes of Daytime Fatigue and Somnolence	270
8.3.4	Subacute Myelo-Optic Neuropathy	246	10.4	Abnormal Movements in Sleep (Parasomnias)	271
8.3.5	Other Autoimmune Diseases	246			
9	Epilepsy and Its Differential Diagnosis	247	11	Polyradiculopathy and Polyneuropathy	273
9.1	Fundamentals	249	11.1	Fundamentals	275
9.1.1	Classification of the Epilepsies	249	11.2	Polyradiculitis	275
9.1.2	Practical Clinical Management of a Suspected Epileptic Seizure	250	11.2.1	Guillain-Barré Syndrome	275
9.2	Generalized Seizures	254	11.2.2	Chronic Inflammatory Demyelinating (Recurrent) Polyneuropathy	277
9.2.1	Tonic-Clonic Seizures (Earlier Term: "Grand Mal Epilepsy")	254	11.2.3	Cranial Polyradiculitis	277
9.2.2	Absences (Earlier Term: "Petit Mal Epilepsy")	256	11.2.4	Polyradiculitis of the Cauda Equina	278
9.2.3	Atypical Absences and Other Types of Epilepsy in Childhood	256	11.3	Polyneuropathy	278
9.3	Focal (Partial) Seizures	256	11.3.1	Particular Etiologic Types of Polyneuropathy	278
9.3.1	Focal Seizures without Altered Consciousness	256	12	Diseases of the Cranial Nerves	285
9.3.2	Focal Seizures with Altered Consciousness (Earlier Term, "Complex Partial Seizures;" Current Term, "Dyscognitive Seizures")	257	12.1	Disturbances of Smell (Olfactory Nerve)	287
9.4	Status Epilepticus	260	12.2	Neurologic Disturbances of Vision (Optic Nerve)	287
9.5	Episodic Neurologic Disturbances of Nonepileptic Origin	260	12.2.1	Visual Field Defects	287
9.5.1	Nonepileptic Psychogenic Seizures	260	12.2.2	Impairment of Visual Acuity	289
9.5.2	Episodic Disturbances with Brief Impairment of Consciousness and Falling	261	12.3	Disturbances of Ocular and Pupillary Motility	290
9.5.3	Episodic Falling without Impairment of Consciousness	263	12.3.1	The General Principles of Eye Movements	291
9.5.4	Episodic Impairment of Consciousness without Falling	263	12.3.2	Nystagmus	291
9.5.5	Episodic Movement Disorders without Impairment of Consciousness	263	12.3.3	Supranuclear Oculomotor Disturbances	295
9.5.6	Episodic Impairment of Memory and Confusion	264	12.3.4	Lesions of the Nerves to the Eye Muscles and Their Brainstem Nuclei	298
10	Sleep and Its Abnormalities	265	12.3.5	Ptosis	300
10.1	Shortened Sleep Duration and Abnormal Sleep-Wake Rhythm	267	12.3.6	Pupillary Disturbances	301
10.2	Insomnia	267	12.4	Lesions of the Trigeminal Nerve	303
10.2.1	General Principles	267	12.5	Lesions of the Facial Nerve	303
10.2.2	Restless Legs Syndrome	267	12.5.1	Topical Classification of Facial Palsy	304
10.3	Hypersomnia and Excessive Daytime Somnolence	268	12.5.2	Etiologic Classification of Facial Palsy	305
10.3.1	Sleep Apnea Syndrome	268	12.6	Disturbances of Hearing and Balance: Vertigo	308
10.3.2	Narcolepsy-Cataplexy Syndrome	269	12.6.1	Neurologic Disturbances of Hearing	309
10.3.3	Kleine-Levin-Critchley Syndrome	270	12.6.2	Disequilibrium and Vertigo	311
			12.7	Lesions of the Glossopharyngeal and Vagus Nerves	315
			12.8	Lesions of the Accessory Nerve	315
			12.9	Lesions of the Hypoglossal Nerve	316

12.10	Multiple Cranial Nerve Deficits	317	14.5	Pain in the Trunk and Back	385
13	Diseases of the Spinal Nerve Roots and Peripheral Nerves	319	14.5.1	Thoracic and Abdominal Wall Pain	385
13.1	Radicular Syndromes	321	14.5.2	Back Pain	385
13.1.1	Overview	321	14.5.3	Groin Pain	387
13.1.2	Radicular Syndromes due to Intervertebral Disk Herniation	323	14.6	Leg Pain	387
13.1.3	Radicular Syndromes due to Spinal Stenosis	329	14.7	Pseudoradicular Pain	388
13.1.4	Radicular Syndromes due to Space-Occupying Lesions	330	15	Diseases of Muscle (Myopathies)	389
13.2	Peripheral Nerve Lesions	331	15.1	Structure and Function of Muscle	391
13.2.1	Overview	331	15.1.1	Microscopic Anatomy of Muscle	391
13.2.2	Diseases of the Brachial Plexus	333	15.1.2	Physiology of Muscle Contraction	391
13.2.3	Diseases of the Peripheral Nerves of the Upper Limbs	338	15.1.3	Impulse Transmission at the Motor End Plate and Impulse Conduction in the Muscle Fiber	391
13.2.4	Diseases of the Nerves of the Trunk	348	15.2	General Symptomatology	392
13.2.5	Diseases of the Lumbosacral Plexus	350	15.3	Muscular Dystrophies	393
13.2.6	Diseases of the Peripheral Nerves of the Lower Limbs	351	15.3.1	Hereditary Muscular Dystrophies of X-chromosomal Inheritance—Dystrophinopathies	396
14	Painful Syndromes	361	15.3.2	Hereditary Muscular Dystrophies of Autosomal Inheritance	398
14.1	Fundamentals	363	15.3.3	Rarer Types of Muscular Dystrophy	400
14.1.1	The Generation and Perception of Pain	363	15.4	Myotonic Syndromes and Periodic Paralysis Syndromes	400
14.1.2	General Aspects of the Clinical History in Patients with Pain	363	15.4.1	Diseases Mainly Causing Myotonia	400
14.2	Painful Syndromes of the Head and Neck	364	15.4.2	Diseases Causing Periodic Paralysis	402
14.2.1	IHS Classification of Headache	364	15.5	Metabolic Myopathies	402
14.2.2	Approach to the Patient with Headache	365	15.5.1	Acute Rhabdomyolysis	402
14.2.3	The Main Types of Primary Headache	366	15.5.2	Mitochondrial Encephalomyopathies	403
14.2.4	The Main Types of Secondary Headache	373	15.6	Myositis	404
14.3	Painful Syndromes of the Face	379	15.6.1	Polymyositis and Dermatomyositis	404
14.3.1	Neuralgias	379	15.7	Other Diseases Affecting Muscle	406
14.3.2	Pain in the Face Caused by Diseases of the Teeth and Jaws	381	15.7.1	Myopathies due to Systemic Disease	406
14.3.3	Atypical Facial Pain	381	15.7.2	Congenital Myopathies	406
14.3.4	Further Types of Facial Pain	381	15.8	Disturbances of Neuromuscular Transmission—Myasthenic Syndromes	406
14.3.5	Differential Diagnosis of Headache and Facial Pain	381	15.8.1	Myasthenia Gravis	407
14.4	Painful Shoulder–Arm Syndromes	381	15.8.2	Lambert–Eaton Syndrome	410
14.4.1	Spondylogenic (Cervicogenic) Shoulder and Arm Pain	382	15.8.3	Rare Myasthenia-like Syndromes	410
14.4.2	Degenerative and Rheumatic Shoulder and Arm Pain	382	16	Diseases of the Autonomic Nervous System	411
14.4.3	Neurogenic Arm Pain	384	16.1	Anatomy	413
14.4.4	Vasogenic Arm Pain	384	16.1.1	Sympathetic Nervous System	413
14.4.5	Arm Pain of Overuse	385	16.1.2	Parasympathetic Nervous System	413
14.4.6	Other Types of Arm Pain	385	16.2	Normal and Pathologic Function of the Autonomic Nervous System	416

16.2.1 Sweating	416
16.2.2 Bladder, Bowel, and Sexual Function	416
16.2.3 The Cervical Sympathetic Pathway and Horner Syndrome	419
16.2.4 Generalized Autonomic Dysfunction	419
<hr/>	
Index	421