

CONTENTS

1	Developmental Anatomy and Physiology of the Central Nervous System From Birth to Two Years of Age	1
	NEONATAL STAGE	1
	THREE MONTHS	6
	SIX MONTHS	12
	NINE MONTHS	16
	TWELVE MONTHS	21
	EIGHTEEN MONTHS	25
	TWENTY-FOUR MONTHS	29
	Comment	34
	Physical Measurements	35
	References	37
2	Examination	39
	History	39
	General Physical Examination	40
	Assessment of the Developmental Age	42
	Neurological Examination	52
	SPECIAL SENSES	52
	CRANIAL NERVES	55
	MOTOR SYSTEM	57
	SENSORY SYSTEM	58
	COORDINATION	59
	STRETCH AND SUPERFICIAL REFLEXES	59
	TRANSITORY REFLEXES, CHARACTERISTIC OF INFANCY	60
	Special Tests	63
	The Autonomic Nervous System	64
	References	65
3	Birth Injuries	66
	Intracranial Birth Injuries	66
	SUBDURAL HEMORRHAGE	67
	MASSIVE INTRACEREBRAL HEMORRHAGE	68
	SUBARACHNOID HEMORRHAGE	72
	INTRAVENTRICULAR HEMORRHAGE	73
	SUBPENDYMAL HEMORRHAGE	74
	MULTIPLE SMALL CEREBRAL HEMORRHAGES	75
	Birth Injury to the Cranial Bones	76
	Asphyxia at Birth	77

Birth Injury to the Spinal Cord	79
Birth Injury to the Brachial Plexus and Peripheral Nerves	80
BIRTH INJURY TO THE FACIAL NERVE	80
BIRTH INJURY TO THE BRACHIAL PLEXUS	82
PHRENIC NERVE PARALYSIS	83
References	85
4 Vascular Disorders Affecting the Central Nervous System	87
Cerebral Hemorrhage	87
Head Trauma	87
SUBDURAL HEMATOMA	88
EPIDURAL HEMORRHAGE	91
MASSIVE INTRACEREBRAL HEMORRHAGE	91
BRAIN CONCUSSION	92
Cerebral Hemorrhage Associated with Various Bleeding Diathesis	92
CEREBRAL HEMORRHAGE IN BLOOD DYSCRASIA	94
HEMORRHAGIC DISEASE OF THE NEWBORN	95
Cerebral Hemorrhages Complicating Infections and Intoxications	96
Cerebral Hemorrhage within a Neoplasm	97
Cerebral Thrombosis	97
ANATOMICAL AND PHYSIOLOGIC CONSIDERATION OF CEREBRAL VENOUS CIRCULATION	98
THROMBOSIS OF CEREBRAL VEINS	99
THROMBOSIS OF THE DURAL SINUSES	100
THROMBOSIS OF THE CEREBRAL ARTERIES	104
THROMBOSIS OF MULTIPLE SMALL CEREBRAL ARTERIES	104
Cerebral Embolism	105
Involvement of the Central Nervous System Secondary to Intrinsic Pathology of the Blood Vessels	106
PERIARTERITIS NODOSA	107
ACUTE DISSEMINATED LUPUS ERYTHEMATOSUS	107
Aneurysms and Arteriovenous Malformations of the Central Nervous System	108
INTRACRANIAL ANEURYSMS	108
ARTERIOVENOUS MALFORMATIONS	109
Cerebral Hemorrhage of Undetermined Cause	111
References	112
5 Encephalitis and Encephalomyelitis	116
Purulent Encephalitis or Encephalomyelitis and Brain Abscess Caused by Bacteria	116
BRAIN ABSCESS, LOCALIZED AND DIFFUSE ENCEPHALITIS	116
Meningoencephalitis and Encephalomyelitis Caused by a Fungus	119

TOXOPLASMOSIS	119
HISTOPLASMOSIS	123
TORULOSIS	124
Congenital Syphilis and Neurosyphilis	127
Encephalitis and Encephalomyelitis Caused by Viruses	130
WESTERN EQUINE ENCEPHALOMYELITIS	130
EASTERN EQUINE ENCEPHALOMYELITIS	132
ST. LOUIS ENCEPHALOMYELITIS	135
JAPANESE B. ENCEPHALOMYELITIS	136
VENEZUELAN EQUINE ENCEPHALOMYELITIS	137
RUSSIAN SPRING-SUMMER ENCEPHALITIS	137
AUSTRALIAN X ENCEPHALITIS	138
HERPES SIMPLEX ENCEPHALITIS AND MENINGOENCEPHALITIS	138
HERPES ZOSTER ENCEPHALITIS	139
RABIES ENCEPHALITIS	139
MUMPS MENINGITIS AND MENINGOENCEPHALOMYELITIS	140
POLIOMYELITIS	141
CYTOMEGALIC INCLUSION BODY DISEASE	145
Postinfectious and Postvaccinal Encephalomyelitis	147
ACUTE ENCEPHALOMYELITIS ASSOCIATED WITH MEASLES	147
ACUTE ENCEPHALOMYELITIS FOLLOWING RUBELLA	148
ACUTE ENCEPHALOMYELITIS COMPLICATING SMALLPOX	149
ACUTE ENCEPHALOMYELITIS COMPLICATING CHICKENPOX	149
ACUTE POSTVACCINAL ENCEPHALOMYELITIS	149
Involvement of the Central Nervous System During the Course of Certain Bacterial Diseases	150
COMPLICATIONS OF THE CENTRAL NERVOUS SYSTEM IN THE COURSE OF PERTUSIS	151
INVOLVEMENT OF THE CENTRAL NERVOUS SYSTEM COMPLICATING SCARLET FEVER	151
CEREBRAL COMPLICATIONS OF INFLUENZA AND NONSUPPURATIVE PNEUMONIA	152
Encephalitis or Encephalomyelitis of Obscure Etiology	152
SYDENHAM'S CHOREA	152
POLYRADICULONEUROPATHY OR GUILLAIN-BARRÉ'S SYNDROME	153
References	154

6	Meningitis and Suppurative Lesions About the Meninges	159
	Purulent Meningitis	159
	Tuberculous Meningitis	166
	Acute Viral Meningitis	170
	LYMPHOCYTIC CHORIOMENINGITIS	172
	MUMPS MENINGITIS	172
	Aseptic Meningitis	172
	Suppurative Lesions about the Meninges	173
	EPIDURAL ABSCESS	173

PETROSITIS AND GRADENIGO SYNDROME	174
SUBDURAL EMPYEMA	174
SUBDURAL EFFUSION	175
References	175

7

Dysraphic States and Various Abnormalities of the Skull and Spine	177
Dysraphic States Involving the Spine and Cranium	177
SPINAL DYSRAPHISM	178
CRANIAL DYSRAPHISM	181
Arnold-Chiari Malformation	186
Craniosostenosis	188
ACROCEPHALOSYNDACTYLY	192
Microcephaly	193
"GENETIC" MICROCEPHALY	193
ACQUIRED OR SECONDARY MICROCEPHALY	194
Less Common Cranial Abnormalities	196
OCULAR HYPERTELORISM	196
CLEIDOCRANIAL DYSOSTOSIS	196
KLIPPEL-FEIL SYNDROME	196
References	197

8

Various Conditions Underlying Cerebral Palsy	199
Congenital Malformations of Various Cerebral Structures	199
MICROPOLYGYRIA	200
PACHYGYRIA	201
RARE FORMS OF CEREBRAL MALFORMATION	203
CEREBELLAR MALFORMATIONS	205
HYDROMYELIA AND SYRINGOMYELIA	206
CLINICAL MANIFESTATIONS OF THE CEREBRAL MALFORMATIONS	207
Neurological Abnormalities of Infants Following Maternal Rubella in the First Trimester	208
Destructive Brain Lesions Occurring During Prenatal, Intranatal or Early Postnatal Life	210
PORENCEPHALY	210
ULEGYRIA	211
CYSTIC DEGENERATION OF THE WHITE MATTER	213
GROSS, LOCALIZED CEREBRAL SCLEROSIS	213
STATUS MARMORATUS	213
CLINICAL MANIFESTATIONS OF DESTRUCTIVE BRAIN LESIONS	213
Descriptive Clinical Entities Such as Hemiplegia, Cerebral Diplegia, Paraplegia and Monoplegia	214
INFANTILE HEMIPLEGIA	215
CEREBRAL DIPLEGIA	217
BILATERAL HEMIPLEGIA	221
QUADRIPLEGIA	221

PARAPLEGIA.....	221
MONOPLEGIA.....	221
References.....	222

9

Hydrocephalus	223
References.....	233

10

Epilepsy	235
Classification of Epilepsy.....	235
Recurrent Epileptic Attacks Associated with a Demonstrable Organic Brain Lesion.....	235
EPILEPTIC MANIFESTATIONS IN PROGRESSIVE CEREBRAL LESIONS.....	235
EPILEPTIC ATTACKS ASSOCIATED WITH NONPROGRESSIVE CEREBRAL LESIONS.....	236
Recurrent Epileptic Attacks not Associated with a Demonstrable Organic Brain Lesion.....	236
Transient Epileptic Attacks During an Acute Illness Affecting the Central Nervous System or in a Generalized Disturbance of Metabolism.....	236
Incidence of Seizures During Infancy.....	239
Plan of Investigation of Cerebral Seizures.....	239
Clinical Forms of the more Commonly Encountered Epileptic Attacks.....	240
GRAND MAL SEIZURES.....	240
PETIT MAL.....	241
PETIT MAL VARIANT.....	242
FOCAL EPILEPTIC ATTACKS.....	244
JACKSONIAN CONVULSIONS.....	245
INFANTILE SPASMS (MASSIVE SPASMS).....	245
MYOCLONIC SEIZURES.....	246
NEONATAL CONVULSIONS.....	247
MYOCLONUS.....	247
PYKNOLEPSY.....	247
Diagnosis of Epilepsy.....	248
Treatment and General Management.....	248
Effects of Frequently Recurring Epileptic Attacks on the Development of the Infant.....	249
References.....	253

11

Mental Deficiency and Certain Disorders Leading to Retardation of Development	254
UNDIFFERENTIATED MENTAL DEFICIENCY.....	255
MONGOLISM.....	257
FAMILIAL DYSAUTONOMIA.....	261

DEVELOPMENTAL SPEECH DISORDERS	262
References	266

12

Exogenous and Endogenous Intoxications Affecting the Nervous System	267
LEAD INTOXICATION	267
ACRODYNIA (PINK DISEASE)	271
TOXIC EFFECTS OF ANAESTHETICS AND HYPNOTICS	272
CARBON MONOXIDE INTOXICATION	273
NEUROLOGIC ABNORMALITIES ENCOUNTERED IN INFANTS WHO SUFFER FROM RETROLENTAL FIBROPLASIA	275
PORPHYRIA	276
INVOLVEMENT OF THE NERVOUS SYSTEM IN BLOOD INCOMPATIBILITY	277
KERNICTERUS	278
References	282

13

Neurocutaneous Dysgeneses	285
STURGE-WEBER DISEASE	285
TUBEROUS SCLEROSIS	287
VON HIPPEL-LINDAU DISEASE	291
NEUROFIBROMATOSIS (VON RECKLINGHAUSEN'S DISEASE)	292
References	294

14

Neoplasms and Granulomatous Conditions Affecting the Nervous System	296
Cerebral Neoplasms	296
GENERAL FEATURES OF CEREBRAL NEOPLASM IN INFANCY	296
MEDULLOBLASTOMA	301
ASTROCYTOMA	301
EPENDYMOMA	302
TUMORS OF THE CHOROID PLEXUS	303
CRANIOPHARYNGIOMA	304
PINEALOMA	305
GLIOMA OF THE OPTIC NERVE	306
GLIOBLASTOMA MULTIFORME	307
MENINGIOMA	307
VASCULAR TUMORS	308
INTRACRANIAL METASTATIC TUMORS	308
Intraspinal Neoplasms	308
INTRASPINAL NEOPLASMS OF DEVELOPMENTAL ORIGIN	311
METASTASES OF MALIGNANT NEOPLASMS INVOLVING THE SPINAL CORD OR NERVE ROOTS	312

Histiocytic Reticuloendotheliosis 315
 HAND-SCHÜLLER-CHRISTIAN SYNDROME 317
 References 319

15

Metabolic and Endocrine Disorders Affecting the Nervous System 321
 Disorders of the Nervous System Associated with Abnormal Lipid Metabolism 321
 AMAUROTIC FAMILIAL IDIOCY OR CEREBROMACULAR DEGENERATION 321
 NIEMANN-PICK'S DISEASE 325
 GAUCHER'S DISEASE 326
 GARGOYLISM 327
 Neurological Involvement in Diseases of Carbohydrate Metabolism 331
 GLYCOGEN STORAGE DISEASE OF THE LIVER (VON GIERKE'S DISEASE) 331
 GLYCOGEN STORAGE DISEASE OF THE HEART AND SKELETAL MUSCLES 332
 GALACTOSEMIA 333
 DIABETES MELLITUS 334
 HYPOGLYCEMIC STATES 335
 Disorders of the Central Nervous System Associated with Disturbed Protein Metabolism 335
 PHENYLPYRUVIC OLIGOPHRENIA 335
 Disorders of the Nervous System Associated with Vitamin Deficiency 339
 PELLAGRA 339
 NEUROPATHY DUE TO THIAMINE DEFICIENCY (BERI-BERI) 340
 RICKETS 341
 NEUROLOGICAL COMPLICATIONS IN CONDITIONS KNOWN AS CELIAC SYNDROME 342
 Neurological Abnormalities Associated with Endocrine Disorders 343
 CRETINISM 343
 HYPOPITUITARISM 346
 DIABETES INSIPIDUS 347
 LAURENCE-BIEDL-MOON SYNDROME 348
 ALDOSTERONISM 350
 HYPERPARATHYROIDISM 351
 HYPOPARATHYROIDISM 352
 TETANY 353
 References 354

16

Progressive Degenerations of the Central Nervous System 357
 PROGRESSIVE INFANTILE MUSCULAR ATROPHY, WERDNIG-HOFFMAN DISEASE 357
 DIFFUSE CEREBRAL SCLEROSIS 359
 HEREDITARY CEREBELLAR AND SPINOCEREBELLAR ATAXIAS 363
 References 364

17

Primary Diseases of the Muscular System	366
Progressive Muscular Dystrophy.....	367
RAPIDLY PROGRESSIVE GENERALIZED MUSCULAR DYSTROPHY.....	367
PREDOMINANTLY LOCALIZED FORMS OF MUSCULAR DYSTROPHY....	370
Myotonic Muscular Dystrophies.....	371
DYSTROPHIA MYOTONICA.....	371
MYOTONIA CONGENITA (THOMSEN).....	371
PARAMYOTONIA CONGENITA.....	372
Congenital Muscular Hypoplasia, Oppenheim-Turner's Syndrome.	372
Polymyositis and Dermatomyositis.....	374
ACUTE POLYMYOSITIS.....	375
DERMATOMYOSITIS.....	375
Arthrogryposis Multiplex Congenita.....	376
Myasthenia Gravis.....	377
Congenital Absence of Isolated Muscles.....	380
References.....	380
 Index.....	 383