

Eva L. Feldman
Wolfgang Grisold
James W. Russell
Wolfgang N. Löscher

Atlas of Neuromuscular Diseases

A Practical Guideline

Second Edition

 Springer

Eva L. Feldman • Wolfgang Grisold
James W. Russell • Wolfgang N. Löscher

Atlas of Neuromuscular Diseases

A Practical Guideline

Second Edition



Eva L. Feldman
A. Alfred Taubman Med. Res. Inst.
Ann Arbor, MI
USA

Wolfgang Grisold
Department of Neurology
Kaiser-Franz-Josef-Spital
Wien
Austria

James W. Russell
Professor, Department of Neurology
and Veterans Administration
Maryland Healthcare System
Neuromuscular Division Head
Director-Peripheral Neuropathy Center
University of Maryland
Baltimore, MD
USA

Wolfgang N. Löscher
Division of Neurology
Department of Neurology and Neurosurgery
Medical University Innsbruck
Innsbruck
Tirol
Austria

ISBN 978-3-7091-1604-3 ISBN 978-3-7091-1605-0 (eBook)
DOI 10.1007/978-3-7091-1605-0
Springer Heidelberg Dordrecht London New York

Library of Congress Control Number: 2014941100

© Springer-Verlag Wien 2014

This work is subject to copyright. All rights are reserved by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed. Exempted from this legal reservation are brief excerpts in connection with reviews or scholarly analysis or material supplied specifically for the purpose of being entered and executed on a computer system, for exclusive use by the purchaser of the work. Duplication of this publication or parts thereof is permitted only under the provisions of the Copyright Law of the Publisher's location, in its current version, and permission for use must always be obtained from Springer. Permissions for use may be obtained through RightsLink at the Copyright Clearance Center. Violations are liable to prosecution under the respective Copyright Law.

The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.

While the advice and information in this book are believed to be true and accurate at the date of publication, neither the authors nor the editors nor the publisher can accept any legal responsibility for any errors or omissions that may be made. The publisher makes no warranty, express or implied, with respect to the material contained herein.

Printed on acid-free paper

Springer is part of Springer Science+Business Media (www.springer.com)

Atlas of Neuromuscular Diseases

This book is dedicated to Professors P.K. Thomas (London, UK) and John Griffin (Baltimore, MD, USA), our mentors in neuromuscular diseases and to our families whose help and support made this book possible.

Acknowledgments

A book of this type requires many contributors and the authors are grateful for the expert assistance we have received from our colleagues. Some colleagues are acknowledged as co-authors of specific chapters.

Mrs. Jeanette Schulz has been responsible for art and cartoons.

We would also like to acknowledge Dr. Barbara Mechtler Horvath, a radiologist with a special interest in MRI studies, for her expert input on the use of MRI in the neuromuscular clinic. Dr. Stefan Meng, a radiologist with a background in neuroanatomy, provided valuable clinical input on the utility of ultrasound in the neuromuscular clinic. We appreciate that he assists us in correlating ultrasound imaging with clinical presentation and electrophysiological assessments.

We are grateful to the members of the Neuromuscular Divisions of the Kaiser Franz Josef Hospital in Vienna, especially Drs. Peter Hitzemberger, Elisabeth Lindeck Pozza and Vera Wolhgenannt, the Department of Neurology, Innsbruck Medical University, especially Dr. Julia Wanschitz, University of Michigan, especially Drs. Brian Callaghan, Stephen Goutman and Ann Little, the University of Maryland, especially Drs. Justin Kwan, Charlene Hafer-Macko, Neil Porter, and Lindsay Zilliox. Their assistance in expanding our clinical neuromuscular knowledge and sharing their expertise on EMG, neuropathology, genetics and imaging studies have increased the quality of this book. We appreciate Jane Russell for assistance with copy-editing the section on Muscle Diseases.

Finally, we would like to thank Dr. Mila Blaivas, Dr. Andrea Vass, Ms. Judy Boldt, Ms. Claudia Steffek and Mrs. Lehnhart and Mr. Bachem from Springer for images, expert secretarial assistance and editing. Their assistance in completion of this book is deeply appreciated.

Collaborators

Rudy J. Castellani, MD Professor of Pathology, Director of Neuropathology, Baltimore, MD, USA

Hsinlin Thomas Cheng, MD, PhD Department of Neurology, Massachusetts General Hospital, Harvard Medical School, Boston, MA, USA

B. Jane Distad, MD Associate Professor of Neurology, Seattle, WA

Eva L. Feldman, MD, PhD, FAAN, FANA Department of Neurology, University of Michigan, Ann Arbor, MI, USA

Wolfgang Grisold Department of Neurology, Kaiser-Franz-Josef-Spital, Wien, Austria

Wolfgang N. Löscher Klinik für Neurologie, Universitätsklinikum Innsbruck, Innsbruck, Austria

Anne Louise Oaklander, MD, PhD Department of Neurology, Massachusetts General Hospital, Harvard Medical School, Boston, MA, USA

T. Paternostro-Sluga Abteilungsvorständin Physikalische Medizin und Rehabilitation, Donauespital, Vienna, Austria

M. Quittan Abteilungsvorstand für Physikalische Medizin und Rehabilitation, Kaiser-Franz-Josef-Spital, Vienna, Austria

James W. Russell, MBChB, MS, FRCP, FACP Professor, Department of Neurology and Veterans Administration Maryland Healthcare System, Neuromuscular Division Head, Director-Peripheral Neuropathy Center, University of Maryland, Baltimore, MD, USA

R. Schmidhammer Millesi Center, Wiener Privatklinik, Vienna, Austria

Walter Struhal, MD Department for Neurology and Psychiatry, General Hospital of the City of Linz, Linz, Austria

M.D. Weiss, MD, FAAN, FANA Professor, Department of Neurology, Director, Division of Neuromuscular Diseases, University of Washington Medical Center, Seattle, WA

Contents

1	Tools	1
1.1	New Developments in Neuromuscular Disease	1
1.2	The Patient with Neuromuscular Disease	2
1.3	History and General Physical Examination	3
1.4	Neuromuscular Clinical Phenomenology	4
1.4.1	Motor Function	4
1.4.2	Abnormal Muscle Movements	5
1.4.3	Reflex Testing	7
1.4.4	Muscle Tone	8
1.4.5	Sensory Symptoms	8
1.5	Sensory Qualities	8
1.5.1	Myalgia and Pain	9
1.5.2	Neuropathic Pain	10
1.5.3	Autonomic Function	10
1.5.4	Gait, Coordination	11
1.5.5	Clinical Pitfalls	11
1.6	NCV/EMG/Autonomic Testing and Miscellaneous Electrophysiology	12
1.6.1	Motor NCV Studies	12
1.6.2	EMG Techniques	14
1.7	Laboratory Tests	14
1.7.1	Autoimmune Testing in Neuromuscular Transmission and Muscle Disorders	15
1.8	Genetic Testing	16
1.9	Neuroimaging Techniques: MR and Ultrasound	16
1.9.1	Imaging of the Spine and Vertebral Column	16
1.9.2	Imaging Muscle Disease	16
1.9.3	Imaging of Peripheral Nerves	18
1.10	Tissue Diagnosis: Muscle/Nerve/Skin Biopsy	18
1.10.1	Nerve Biopsy	19
1.10.2	Muscle Biopsy	19
1.11	Neuromuscular Approaches to Intervention: Effects of Regional Anesthetic Procedures	19
	References	20
2	Principles of Peripheral Nerve Surgery	23
2.1	Defining the Problem	23
2.2	Timing of Nerve Repair	23
2.3	Restoration of Nerve Continuity	23
2.3.1	End-to-End Coaptation (Direct Nerve Repair)	23
2.3.2	Nerve Grafting	25

2.4	End-to-Side Coaptation	25
2.5	Nerve Transfer	26
2.6	Neurolysis	26
	References	26
3	Principles of Nerve and Muscle Rehabilitation	27
3.1	Principles	27
3.2	Outcome Measurement	27
3.3	Rehabilitation Treatment	28
3.3.1	Exercise and Medical Training	28
3.3.2	Occupational Therapy and Splints	30
3.3.3	Orthoses	30
3.3.4	Neural Plasticity	30
3.3.5	Surgery	31
3.3.6	Physical Modalities	31
3.3.7	Treatment Options for Autonomic Symptoms	32
3.4	Mononeuropathies	32
3.4.1	Median Neuropathy	32
3.4.2	Ulnar Neuropathy	32
3.4.3	Femoral Neuropathy	33
3.4.4	Peroneal Neuropathy	33
3.4.5	Tibial Neuropathy	33
3.4.6	Plexopathies	33
3.5	Polyneuropathies	33
3.6	Myopathies	34
	References	34
4	Chronic Pain in Neuromuscular Disease	37
4.1	Introduction	37
4.2	Clinical Approach and Treatments to Neuropathic Pain	37
4.2.1	Diagnosis	37
4.2.2	Common Patterns of Peripheral Neuropathic Pain	38
4.2.3	Pharmacological Treatments Options	39
4.2.4	Neurosurgical Treatment Options	41
	References	41
5	Cranial Nerve	43
5.1	Introduction	43
5.2	Olfactory Nerve	43
5.3	Optic Nerve	44
5.4	Oculomotor Nerve	45
5.5	Trochlear Nerve	48
5.6	Trigeminal Nerve	49
5.7	Abducens Nerve	54
5.8	Facial Nerve	55
5.9	Acoustic Nerve	58
5.10	Vestibular Nerve	59
5.11	Glossopharyngeal Nerve	60
5.12	Vagus Nerve	61
5.13	Accessory Nerve	62
5.14	Hypoglossal Nerve	64
5.15	Oral Cavity	65
5.15.1	Ventral Part and Closure	66
5.15.2	Middle Part, Oral Cavity and Tongue	66
5.15.3	Posterior Part, Gag and Swallowing	66

5.16	Cranial Nerves and Painful Conditions: A Checklist	67
5.17	Cranial Nerve Examination in Coma	67
5.18	Pupil	68
5.19	Multiple and Combined Oculomotor Nerve Palsies	69
	References	70
6	Radiculopathies	73
6.1	Cervical Radicular Symptoms.	73
6.1.1	Anatomy	73
6.1.2	Symptoms	73
6.1.3	Signs	75
6.1.4	Pathogenesis	75
6.1.5	Diagnosis.	76
6.1.6	Differential Diagnosis	76
6.1.7	Treatment	76
6.1.8	Prognosis.	76
6.2	Thoracic Radicular Nerves	76
6.2.1	Anatomy	77
6.2.2	Symptoms	78
6.2.3	Signs	78
6.2.4	Pathogenesis	78
6.2.5	Diagnosis.	79
6.2.6	Differential Diagnosis	79
6.2.7	Therapy	79
6.2.8	Prognosis.	79
6.3	Lumbar and Sacral Radiculopathy	79
6.3.1	Anatomy	80
6.3.2	Symptoms	80
6.3.3	Signs	81
6.3.4	Pathogenesis	81
6.3.5	Diagnosis.	83
6.3.6	Differential Diagnosis	83
6.3.7	Therapy	83
6.3.8	Prognosis.	84
6.4	Cauda Equina	84
6.4.1	Anatomy	84
6.4.2	Symptoms	85
6.4.3	Signs	85
6.4.4	Pathogenesis	85
6.4.5	Diagnosis.	85
6.4.6	Differential Diagnosis	85
6.4.7	Therapy	85
	References	85
7	Plexopathies	87
7.1	Cervical Plexus and Cervical Spinal Nerves.	87
7.1.1	Anatomy	87
7.1.2	Clinical Picture	87
7.1.3	Symptoms	87
7.1.4	Pathogenesis	87
7.1.5	Diagnosis.	88
7.1.6	Differential Diagnosis	88
7.1.7	Therapy	88

7.2	Brachial Plexus	88
7.2.1	Anatomy	88
7.2.2	Lesions of the Brachial Plexus	89
7.2.3	Symptoms	89
7.2.4	Signs	89
7.2.5	Pathogenesis	90
7.2.6	Diagnosis of Brachial Plexus Lesions	96
7.2.7	Differential Diagnosis	96
7.2.8	Therapy	97
7.2.9	Prognosis	98
7.3	Thoracic Outlet Syndromes	98
7.3.1	True Neurogenic TOS	98
7.3.2	Arterial TOS	98
7.3.3	Venous TOS	99
7.3.4	Disputed Neurogenic TOS	99
7.3.5	Others	99
7.4	Lumbosacral Plexus	99
7.4.1	Anatomy	99
7.4.2	Symptoms	101
7.4.3	Signs	101
7.4.4	Pathogenesis	101
7.4.5	Diagnosis	104
7.4.6	Differential Diagnosis	104
7.4.7	Therapy	104
7.4.8	Prognosis	104
	References	104
8	Mononeuropathies	107
8.1	Introduction	107
8.2	Mononeuropathies: Upper Extremities	107
8.2.1	Axillary Nerve	107
8.2.2	Musculocutaneous Nerve	109
8.2.3	Nerves Around the Elbow	111
8.2.4	Median Nerve	112
8.2.5	Ulnar Nerve	120
8.2.6	Radial Nerve	126
8.2.7	Cutaneous Forearm Nerves	130
8.2.8	Digital Nerves of the Hand	132
8.3	Truncal Mononeuropathies	133
8.3.1	Phrenic Nerve	133
8.3.2	Dorsal Scapular Nerve	136
8.3.3	Suprascapular Nerve	136
8.3.4	Subscapular Nerve (Inferior Scapular Nerve)	137
8.3.5	Long Thoracic Nerve	138
8.3.6	Thoracodorsal Nerve	139
8.3.7	Innervation of the Shoulder	140
8.3.8	Pectoral Nerve	143
8.3.9	Thoracic Spinal Nerves	145
8.3.10	Intercostal Nerves	145
8.3.11	Intercostobrachial Nerve	146
8.3.12	Around the Breast	147

8.3.13	Abdominal Walls and Their Innervation	147
8.3.14	Iliohypogastric Nerve	151
8.3.15	Ilioinguinal Nerve	151
8.3.16	Genitofemoral Nerve	153
8.3.17	Superior and Inferior Gluteal Nerves	154
8.3.18	Pudendal Nerve	155
8.4	Mononeuropathies: Lower Extremities	157
8.4.1	Obturator Nerve	157
8.4.2	Neurology and the Hip	158
8.4.3	Femoral Nerve	159
8.4.4	Saphenous Nerve	161
8.4.5	Lateral Femoral Cutaneous Nerve	161
8.4.6	Posterior Cutaneous Femoral Nerve	163
8.4.7	Sciatic Nerve	164
8.4.8	Around the Knee	168
8.4.9	Peroneal Nerve	169
8.4.10	Tibial Nerve (Posterior Tibial Nerve)	172
8.4.11	Sural Nerve	176
8.4.12	Posterior Tarsal Tunnel Syndrome	177
8.4.13	Anterior Tarsal Tunnel Syndrome	178
8.4.14	Interdigital Neuroma and Neuritis	179
8.4.15	Nerves of the Foot	180
8.4.16	Peripheral Nerve Tumors	182
	References	187
9	Polyneuropathies	191
9.1	Introduction	191
9.1.1	Anatomical Distribution	191
9.1.2	Clinical Syndrome	191
9.2	Metabolic Diseases	193
9.2.1	Diabetic Distal Symmetric Polyneuropathy	193
9.2.2	Diabetic Autonomic Neuropathy	195
9.2.3	Diabetic Mononeuritis Multiplex and Diabetic Polyradiculopathy (Amyotrophy)	195
9.2.4	Distal Symmetric Polyneuropathy of Renal Disease	196
9.3	Systemic Diseases	197
9.3.1	Amyloid Neuropathies	197
9.4	Neuropathies Associated with Paraproteinemias	198
9.4.1	Multiple Myeloma Neuropathy	198
9.4.2	Monoclonal Gammopathy of Undetermined Significance (MGUS)	199
9.4.3	Waldenström's Macroglobulinemia	199
9.4.4	Osteosclerotic Myeloma (POEMS Syndrome)	199
9.4.5	Vasculitic Neuropathy, Nonsystemic	200
9.4.6	Vasculitic Neuropathy, Systemic	200
9.4.7	Critical Illness Neuropathy (CIP)	202
9.5	Infectious Neuropathies	203
9.5.1	Human Immunodeficiency Virus-1 Neuropathy	203
9.5.2	Herpes Zoster Neuropathy	204
9.5.3	Lyme Disease (Neuroborreliosis)	204
9.5.4	Leprosy	206

9.6	Inflammatory Neuropathies.	207
9.6.1	Acute Inflammatory Demyelinating Polyneuropathy (AIDP, Guillain–Barre Syndrome)	207
9.6.2	Acute Motor Axonal Neuropathy	207
9.6.3	Acute Motor and Sensory Axonal Neuropathy	209
9.6.4	Miller Fisher Syndrome (MFS).	209
9.6.5	Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)	210
9.6.6	Demyelinating Neuropathy Associated with Anti-MAG Antibodies.	212
9.6.7	Multifocal Motor Neuropathy (MMN)	212
9.7	Nutritional Neuropathies.	213
9.7.1	Cobalamin Neuropathy	213
9.7.2	Post-gastroplasty Neuropathy	213
9.7.3	Pyridoxine Neuropathy	214
9.7.4	Strachan’s Syndrome.	214
9.7.5	Thiamine Neuropathy	214
9.7.6	Tocopherol Neuropathy.	215
9.8	Drugs, Industrial Agents, and Metals	215
9.8.1	Alcohol Polyneuropathy	215
9.8.2	Other Drug-Induced Neuropathies	216
9.8.3	Toxic Neuropathies: Industrial Agents	218
9.8.4	Toxic Neuropathies: Metals	219
9.9	Hereditary Neuropathies	220
9.9.1	Hereditary Motor and Sensory Neuropathies: Charcot-Marie-Tooth Disease	220
9.9.2	Other Hereditary Motor and Sensory Neuropathies	223
9.9.3	Porphyria.	227
9.10	Cancer and Neuropathy	227
9.10.1	Paraneoplastic Neuropathies	227
9.10.2	Motor Neuropathy or Motor Neuron Disease Syndrome.	230
9.10.3	Neuropathies and Neuromyopathies	230
9.10.4	Neuropathies in Lymphoma and Leukemia	230
9.10.5	Neoplastic Neuropathies	231
9.10.6	Polyneuropathy and Chemotherapy	232
	References	234
10	Neuromuscular Transmission: Endplate Disorders	235
10.1	Myasthenia Gravis.	235
10.1.1	Epidemiology	235
10.1.2	Anatomy and Pathophysiology	235
10.1.3	Symptoms	235
10.1.4	Signs	235
10.1.5	Myasthenic Crisis	235
10.1.6	Causes	235
10.1.7	Electrophysiology	238
10.1.8	Imaging (MR, CT Scan)	238
10.1.9	Laboratory	238
10.1.10	Diagnosis.	239
10.1.11	Differential Diagnosis	239
10.1.12	Medication and Myasthenia	239
10.1.13	Therapy	239

10.1.14	Myasthenia and Pregnancy	240
10.1.15	Prognosis	241
10.2	Congenital Myasthenic Syndromes	241
10.3	Lambert-Eaton Myasthenic Syndrome (LEMS)	241
10.3.1	Epidemiology	241
10.3.2	Anatomy and Pathophysiology	241
10.3.3	Symptoms	242
10.3.4	Signs	242
10.3.5	Causes	242
10.3.6	Electrophysiology	242
10.3.7	Imaging	242
10.3.8	Laboratory	243
10.3.9	Diagnosis	243
10.3.10	Differential Diagnosis	243
10.3.11	Therapy	243
10.3.12	Prognosis	243
10.4	Botulism	243
10.4.1	Epidemiology	243
10.4.2	Anatomy and Pathophysiology	243
10.4.3	Symptoms	243
10.4.4	Signs	243
10.4.5	Causes	243
10.4.6	Electrophysiology	243
10.4.7	Imaging	244
10.4.8	Laboratory	244
10.4.9	Diagnosis	244
10.4.10	Differential Diagnosis	244
10.4.11	Therapy	244
10.4.12	Prognosis	244
10.5	Neuromyotonia (Isaacs' Syndrome)	244
10.5.1	Epidemiology	244
10.5.2	Anatomy and Pathophysiology	244
10.5.3	Symptoms	244
10.5.4	Signs	244
10.5.5	Causes	244
10.5.6	Electrophysiology	244
10.5.7	Imaging	245
10.5.8	Laboratory	245
10.5.9	Diagnosis	245
10.5.10	Differential Diagnosis	245
10.5.11	Therapy	245
10.5.12	Prognosis	245
	References	245
11	Muscle and Myotonic Diseases	247
11.1	Introduction	247
11.1.1	Electrophysiology	247
11.1.2	Muscle Histology and Immunohistochemistry	247
11.1.3	Regulation of Gene Defects in Muscle	248
11.2	Polymyositis (PM) and Dermatomyositis (DM)	248
11.2.1	Distribution	248
11.2.2	Clinical Syndrome	248

11.2.3	Pathogenesis	248
11.2.4	Diagnosis.	248
11.2.5	Differential Diagnosis	249
11.2.6	Therapy	249
11.2.7	Prognosis.	250
11.3	Inclusion Body Myositis (IBM)	250
11.3.1	Distribution	250
11.3.2	Clinical Description	250
11.3.3	Pathogenesis	250
11.3.4	Diagnosis.	250
11.3.5	Differential Diagnosis	250
11.3.6	Therapy	251
11.3.7	Prognosis.	251
11.4	Immune-Mediated Necrotizing Myopathy (IMNM).	251
11.4.1	Distribution	251
11.4.2	Clinical Syndrome.	251
11.4.3	Pathogenesis	251
11.4.4	Diagnosis.	251
11.4.5	Differential Diagnosis	252
11.4.6	Treatment	252
11.4.7	Prognosis.	252
11.5	Connective Tissue Diseases (CTDs) in “Overlap” Myositis.	252
11.5.1	Distribution/Anatomy	252
11.5.2	Clinical Syndrome.	252
11.5.3	Pathogenesis	252
11.5.4	Diagnosis.	253
11.5.5	Differential Diagnosis	253
11.5.6	Therapy	253
11.5.7	Prognosis.	254
11.6	Viral Myopathies.	254
11.6.1	Distribution/Anatomy	254
11.6.2	Clinical Syndrome.	254
11.6.3	Pathogenesis	254
11.6.4	Diagnosis.	254
11.6.5	Differential Diagnosis	254
11.6.6	Therapy	254
11.6.7	Prognosis.	255
11.7	Duchenne Muscular Dystrophy.	255
11.7.1	Distribution	255
11.7.2	Clinical Syndrome.	255
11.7.3	Pathogenesis	255
11.7.4	Diagnosis.	256
11.7.5	Differential Diagnosis	256
11.7.6	Therapy	256
11.7.7	Prognosis.	257
11.8	Becker Muscular Dystrophy	257
11.8.1	Distribution/Anatomy	257
11.8.2	Clinical Syndrome.	257
11.8.3	Pathogenesis	257
11.8.4	Diagnosis.	257
11.8.5	Differential Diagnosis	257
11.8.6	Therapy	258
11.8.7	Prognosis.	258

11.9	Myotonic Dystrophy (DM).....	258
11.9.1	Distribution/Anatomy.....	258
11.9.2	Clinical Syndrome.....	258
11.9.3	Pathogenesis.....	258
11.9.4	Diagnosis.....	258
11.9.5	Differential Diagnosis.....	259
11.9.6	Therapy.....	259
11.9.7	Prognosis.....	259
11.10	Limb-Girdle Muscular Dystrophy (LGMD).....	259
11.10.1	Distribution.....	259
11.10.2	Clinical Syndrome.....	259
11.10.3	Pathogenesis.....	260
11.10.4	Diagnosis.....	260
11.10.5	Differential Diagnosis.....	261
11.10.6	Therapy.....	261
11.10.7	Prognosis.....	261
11.11	Oculopharyngeal Muscular Dystrophy (OPMD).....	261
11.11.1	Distribution.....	261
11.11.2	Clinical Syndrome.....	261
11.11.3	Pathogenesis.....	261
11.11.4	Diagnosis.....	261
11.11.5	Differential Diagnosis.....	262
11.11.6	Therapy.....	262
11.11.7	Prognosis.....	262
11.12	Facioscapulohumeral Muscular Dystrophy (FSHD).....	262
11.12.1	Distribution.....	262
11.12.2	Clinical Syndrome.....	262
11.12.3	Pathogenesis.....	264
11.12.4	Diagnosis.....	264
11.12.5	Differential Diagnosis.....	264
11.12.6	Therapy.....	264
11.12.7	Prognosis.....	264
11.13	Distal Myopathies.....	264
11.13.1	Distribution.....	264
11.13.2	Clinical Syndrome.....	264
11.13.3	Pathogenesis.....	264
11.13.4	Diagnosis.....	264
11.13.5	Differential Diagnosis.....	265
11.13.6	Therapy.....	265
11.13.7	Prognosis.....	265
11.14	Congenital Myopathies.....	265
11.14.1	Distribution/Anatomy.....	265
11.14.2	Clinical Syndrome.....	265
11.14.3	Pathogenesis.....	266
11.14.4	Diagnosis.....	266
11.14.5	Differential Diagnosis.....	267
11.14.6	Therapy.....	267
11.14.7	Prognosis.....	268
11.15	Mitochondrial Myopathies.....	268
11.15.1	Distribution/Anatomy.....	268
11.15.2	Clinical Syndrome.....	268
11.15.3	Pathogenesis.....	268
11.15.4	Diagnosis.....	268

11.15.5	Differential Diagnosis.....	268
11.15.6	Therapy.....	268
11.15.7	Prognosis.....	268
11.16	Glycogen Storage Diseases.....	269
11.16.1	Distribution.....	269
11.16.2	Clinical Syndrome.....	269
11.16.3	Pathogenesis.....	269
11.16.4	Diagnosis.....	269
11.16.5	Differential Diagnosis.....	271
11.16.6	Therapy.....	271
11.16.7	Prognosis.....	271
11.17	Defects of Fatty Acid Metabolism.....	271
11.17.1	Distribution.....	271
11.17.2	Clinical Syndrome.....	271
11.17.3	Pathogenesis.....	271
11.17.4	Diagnosis.....	272
11.17.5	Differential Diagnosis.....	272
11.17.6	Therapy.....	272
11.17.7	Prognosis.....	272
11.18	Toxic Myopathies.....	272
11.18.1	Distribution/Anatomy.....	272
11.18.2	Clinical Syndrome.....	272
11.18.3	Pathogenesis.....	273
11.18.4	Diagnosis.....	273
11.18.5	Differential Diagnosis.....	273
11.18.6	Therapy.....	273
11.18.7	Prognosis.....	274
11.19	Critical Illness Myopathy.....	274
11.19.1	Distribution/Anatomy.....	274
11.19.2	Clinical Syndrome.....	274
11.19.3	Pathogenesis.....	274
11.19.4	Diagnosis.....	274
11.19.5	Differential Diagnosis.....	274
11.19.6	Therapy.....	274
11.19.7	Prognosis.....	274
11.20	Myopathies Associated with Endocrine/Metabolic Disorders and Carcinoma.....	274
11.20.1	Distribution/Anatomy.....	274
11.20.2	Clinical Syndrome.....	274
11.20.3	Pathogenesis.....	275
11.20.4	Diagnosis.....	275
11.20.5	Differential Diagnosis.....	275
11.20.6	Therapy.....	275
11.20.7	Prognosis.....	275
11.21	Myotonia Congenita.....	275
11.21.1	Distribution/Anatomy.....	275
11.21.2	Clinical Syndrome.....	275
11.21.3	Pathogenesis.....	276
11.21.4	Diagnosis.....	276
11.21.5	Differential Diagnosis.....	276