

Contents

Contributing Authors.....	xiii
Preface	xix
Foreword	xxi

Section 1: Introduction to Concepts and Classifications

1. History and Classification of “Myoclonic” Epilepsies: From Seizures to Syndromes to Diseases	1 <i>Pierre Genton, Joseph Roger, Renzo Guerrini, Marco T. Medina, Michelle Bureau, Charlotte Dravet, and Antonio V. Delgado-Escueta</i>
2. Ontogeny of the Reticular Formation: Its Possible Relation to the Myoclonic Epilepsies	15 <i>Harvey B. Sarnat</i>
3. Pathophysiology of Myoclonic Epilepsies.....	23 <i>Renzo Guerrini, Paolo Bonanni, Lucio Parmeggiani, Mark Hallett, and Hirokazu Oguni</i>
4. Progressive Myoclonus Epilepsies: EPM1, EPM2A, EPM2B.....	47 <i>Elayne M. Chan, Danielle M. Andrade, Silvana Franceschetti and Berge Minassian</i>
5. Myoclonic Status in Nonprogressive Encephalopathies	59 <i>Bernardo Dalla Bernardina, Elena Fontana, and Francesca Darra</i>

Section 2: Myoclonic Epilepsies of Infancy and Childhood

6. Severe Myoclonic Epilepsy in Infancy: Dravet Syndrome	71 <i>Charlotte Dravet, Michelle Bureau, Hirokazu Oguni, Yukio Fukuyama, and Ozlem Cokar</i>
7. Severe Myoclonic Epilepsy in Infancy: Clinical Analysis and Relation to SCN1A Mutations in a Japanese Cohort	103 <i>Hirokazu Oguni, Kitami Hayashi, Makiko Osawa, Yutaka Awaya, Yukio Fukuyama, Goryu Fukuma, Shinichi Hirose, Akihisa Mitsudome, and Sunao Kaneko</i>
8. Myoclonic Seizures in the Context of Generalized Epilepsy with Febrile Seizures Plus (GEFS ⁺).....	119 <i>Michel Baulac, Isabelle Gourfinkel-An, Stephanie Baulac, and Eric Leguern</i>
9. Benign Myoclonic Epilepsy in Infancy	127 <i>Charlotte Dravet and Michelle Bureau</i>

10. Autosomal Recessive Benign Myoclonic Epilepsy of Infancy 139
Federico Zara and Fabrizio A. De Falco
11. Myoclonic–Astatic Epilepsy of Early Childhood—Definition, Course, Nosography, Genetics 147
B. A. Neubauer, A. Hahn, and H. Doose, I. Tuxhorn
12. Idiopathic Myoclonic–Astatic Epilepsy of Early Childhood—Nosology Based on Electrophysiologic and Long-Term Follow-Up Study of Patients 157
Hirokazu Oguni, Kitami Hayashi, Kaoru Imai, Makoto Funatsuka, Masako Sakauchi, Seigo Shirakawa, and Makiko Osawa
13. Myoclonic Absences: The Seizure and the Syndrome 175
Michelle Bureau and Carlo Alberto Tassinari
14. Eyelid Myoclonia and Absence 185
Athanasios Covatis
15. Childhood Absence Epilepsy Evolving to Juvenile Myoclonic Epilepsy: Electroclinical and Genetic Features 197
Marco T. Medina, Reyna M. Durón, María Elisa Alonso, Charlotte Dravet, Lourdes León, Minerva López-Ruiz, Ricardo Ramos-Ramírez, Ignacio Pascual Castroviejo, Karen Weissbecker, B. Westling, Katerina Tanya Perez-Gosiengfiao, Sonia Khan, Gregorio Pineda, Ryoji Morita, Astrid Rasmussen-Almarez, Jaime Ramos Peek, Sergio Córdova, Iris E. Martínez-Juárez, Francisco Rubio-Donnadieu, Adriana Ochoa-Morales, Aurelio Jara-Prado, Julia N. Bailey, Miyabi Tanaka, Dongsheng Bai, Jesús Machado-Salas, and Antonio V. Delgado-Escueta
16. Photosensitivity: Genetics and Clinical Significance 217
B. A. Neubauer, St. Waltz, M. Grothe, A. Hahn, I. Tuxhorn, T. Sander, G. Kurlemann, and Ulrich Stephan

Section 3: Myoclonic Epilepsies of Adolescence and Adulthood

17. Familial Juvenile Myoclonic Epilepsy 227
María Elisa Alonso, Marco T. Medina, Iris E. Martínez-Juárez, Reyna M. Durón, Julia N. Bailey, Minerva López-Ruiz, Ricardo Ramos-Ramírez, Adriana Ochoa-Morales, Aurelio Jara-Prado, Astrid Rasmussen, Lourdes León, Gregorio Pineda, Ignacio Pascual Castroviejo, Sonia Khan, René Silva, Lizardo Mija, Lucio Portilla, Dongsheng Bai, Katerina Tanya Perez-Gosiengfiao, Jesús Machado-Salas and A.V. Delgado-Escueta
18. Genetics of Juvenile Myoclonic Epilepsy: Faulty Components and Faulty Wiring? 245
Martina Durner, Deb Pal, and David Greenberg

19.	Autosomal Dominant Juvenile Myoclonic Epilepsy and <i>GABRA1</i>	255
	<i>Patrick Cossette, Anne Lortie, Michel Vanasse, Jean-Marc Saint-Hilaire, and Guy A. Rouleau</i>	
20.	<i>CLCN2</i> and Idiopathic Generalized Epilepsy	265
	<i>Armin Heils</i>	
21.	Autosomal Dominant Cortical Myoclonus and Epilepsy (ADCME) with Linkage to Chromosome 2p11.1-q12.2	273
	<i>Renzo Guerrini, Lucio Parmeggiani, Carla Marini, Paola Brovedani, and Paolo Bonanni</i>	
22.	Familial Adult Myoclonic Epilepsy (FAME)	281
	<i>Eiichiro Uyama, Ying-Hui Fu, and Louis J. Ptácek</i>	

Section 4: Treatment of Myoclonic Epilepsies

23.	Treatment of Myoclonic Epilepsies in Infancy and Early Childhood	289
	<i>Raman Sankar, James W. Wheless, Charlotte Dravet, Renzo Guerrini, Marco T. Medina, Michelle Bureau, Pierre Genton, and Antonio V. Delgado-Escueta</i>	
24.	Ketogenic Diet in Patients with Dravet Syndrome and Myoclonic Epilepsies in Infancy and Early Childhood	299
	<i>Natalio Fejerman, Roberto Caraballo, and Ricardo Cersosimo</i>	
25.	Treatment of Myoclonic Epilepsies of Childhood, Adolescence, and Adulthood	307
	<i>Marco T. Medina, Iris E. Martínez-Juárez, Reyna M. Durón, Pierre Genton, Renzo Guerrini, Charlotte Dravet, Michelle Bureau, Katerina Tanya Perez-Gosiengfiao, Claudia Amador, Julia N. Bailey, Franz Chaves-Sell, and Antonio V. Delgado-Escueta</i>	
	Index	325