Contents

1 Introduction: Reminiscing on Models and Modeling .................. 1
   Robert B. Denman

Part I Ex Vivo Models

2 Probing Astrocyte Function in Fragile X Syndrome ............... 15
   Shelley Jacobs, Connie Cheng, and Laurie C. Doering

3 Neural Stem Cells .......................................................... 33
   Maija Castrén

4 Fragile X Mental Retardation Protein (FMRP)
   and the Spinal Sensory System ........................................ 41
   Theodore J. Price and Ohannes K. Melemedjian

5 The Role of the Postsynaptic Density in the Pathology
   of the Fragile X Syndrome ............................................ 61
   Stefan Kindler and Hans-Jürgen Kreienkamp

Part II Non-mouse Eukaryote Models

6 Behavior in a Drosophila Model of Fragile X ....................... 83
   Sean M. McBride, Aaron J. Bell, and Thomas A. Jongens

7 Molecular and Genetic Analysis of the Drosophila Model
   of Fragile X Syndrome .................................................. 119
   Charles R. Tessier and Kendal Broadie
8 Fragile X Mental Retardation Protein and Stem Cells ............... 157
   Abrar Qurashi, Xuekun Li, and Peng Jin

9 Manipulating the Fragile X Mental Retardation Proteins in the Frog ............................................................... 165
   Marc-Etienne Huot, Nicolas Bisson, Thomas Moss, and Edouard W. Khandjian

10 Exploring the Zebra Finch Taeniopygia guttata as a Novel Animal Model for the Speech–Language Deficit of Fragile X Syndrome ................................................................. 181
   Claudia Winograd and Stephanie Ceman

Part III Novel Mouse Models

11 Neuroendocrine Alterations in the Fragile X Mouse ................. 201
   Abdeslem El Idrissi, Xin Yan, William L’Amoreaux, W. Ted Brown, and Carl Dobkin

12 Taking STEPs Forward to Understand Fragile X Syndrome ....... 223
   Susan M. Goebel-Goody and Paul J. Lombroso

13 Fmr-1 as an Offspring Genetic and a Maternal Environmental Factor in Neurodevelopmental Disease ............. 243
   Bojana Zupan and Miklos Toth

14 Mouse Models of the Fragile X Premutation and the Fragile X Associated Tremor/Ataxia Syndrome ..................... 255
   Michael R. Hunsaker, Gloria Arque, Robert F. Berman, Rob Willemsen, and Renate K. Hukema

Part IV The Human Model

15 Clinical Aspects of the Fragile X Syndrome .......................... 273
   W. Ted Brown

16 Fragile X Syndrome: A Psychiatric Perspective ...................... 281
   Michael R. Tranfaglia

17 Fragile X Syndrome and Targeted Treatment Trials .................. 297
   Randi Hagerman, Julie Lauterborn, Jacky Au, and Elizabeth Berry-Kravis
<table>
<thead>
<tr>
<th>Chapter</th>
<th>Title</th>
<th>Pages</th>
</tr>
</thead>
<tbody>
<tr>
<td>18</td>
<td>The Fragile X-Associated Tremor Ataxia Syndrome</td>
<td>337</td>
</tr>
<tr>
<td></td>
<td>Flora Tassone and Randi Hagerman</td>
<td></td>
</tr>
<tr>
<td></td>
<td><strong>Part V Missing Models</strong></td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>Vignettes: Models in Absentia</td>
<td>361</td>
</tr>
<tr>
<td></td>
<td>Robert B. Denman</td>
<td></td>
</tr>
<tr>
<td></td>
<td><strong>Index</strong></td>
<td>385</td>
</tr>
</tbody>
</table>