

Contents

<i>Preface</i>	<i>v</i>
<i>Contributors</i>	<i>ix</i>
 Part I: MITOCHONDRIAL DNA REPLICATION, TRANSCRIPTION, TRANSLATION, AND RESPIRATORY COMPLEX ASSEMBLY	
1. Biochemical Isolation of mtDNA Nucleoids from Animal Cells	3
<i>Daniel F. Bogenhagen</i>	
2. Analysis of Mitochondrial DNA by Two-Dimensional Agarose Gel Electrophoresis	15
<i>Aurelio Reyes, Takehiro Yasukawa, Tricia J. Cluett, and Ian J. Holt</i>	
3. Comparative Purification Strategies for <i>Drosophila</i> and Human Mitochondrial DNA Replication Proteins: DNA Polymerase γ and Mitochondrial Single-Stranded DNA-Binding Protein.	37
<i>Marcos T. Oliveira and Laurie S. Kaguni</i>	
4. Functional Analysis of Mutant Mitochondrial DNA Polymerase Proteins Involved in Human Disease	59
<i>Sherine S. L. Chan and William C. Copeland</i>	
5. Preparation of Human Mitochondrial Single-Stranded DNA-Binding Protein.	73
<i>Matthew J. Longley, Leslie A. Smith, and William C. Copeland</i>	
6. Methods for Assessing Binding of Mitochondrial Transcription Factor A (TFAM) to DNA	87
<i>Atsushi Fukuoka and Dongchon Kang</i>	
7. Inducible Expression in Human Cells, Purification, and In Vitro Assays for the Mitochondrial DNA Helicase Twinkle	103
<i>Steffi Goffart and Hans Spelbrink</i>	
8. Purification Strategy for Recombinant Forms of the Human Mitochondrial DNA Helicase	121
<i>Tawn D. Ziebarth and Laurie S. Kaguni</i>	
9. Methods for Studying Mitochondrial Transcription Termination with Isolated Components	127
<i>Paola Loguercio Polosa, Stefania Deceglie, Marina Roberti, Maria Nicola Gadaleta, and Palmiro Cantatore</i>	
10. Oxidative Phosphorylation: Synthesis of Mitochondrially Encoded Proteins and Assembly of Individual Structural Subunits into Functional Holoenzyme Complexes.	143
<i>Scot C. Leary and Florin Sasarman</i>	

Part II: MITOCHONDRIAL DNA DAMAGE AND REPAIR

11. Reactive Oxygen Species Production by Mitochondria 165
Adrian J. Lambert and Martin D. Brand
12. Measuring mtDNA Damage Using a Supercoiling-Sensitive
qPCR Approach 183
Sam W. Chan and Junjian Z. Chen
13. Quantitative Analysis of Oxidized Guanine, 8-Oxoguanine, in Mitochondrial
DNA by Immunofluorescence Method 199
Mizuki Ohno, Sugako Oka, and Yusaku Nakabeppu
14. *In Vitro* Measurement of DNA Base Excision Repair
in Isolated Mitochondria 213
Melissa M. Page and Jeffrey A. Stuart
15. Targeting Repair Proteins to the Mitochondria of Mammalian Cells Through
Stable Transfection, Transient Transfection, Viral Transduction, and
TAT-Mediated Protein Transduction 233
*Christopher A. Koczor, Janet W. Snyder, Inna N. Shokolenko,
Allison W. Dobson, Glenn L. Wilson, and Susan P. LeDoux*
16. Construction and Characterization of a Cell Line Deficient in Repair of
Mitochondrial, but Not Nuclear, Oxidative DNA Damage 251
Sugako Oka, Mizuki Ohno, and Yusaku Nakabeppu

Part III: MITOCHONDRIAL DNA MUTATIONS

17. Mitochondrial DNA Oxidative Damage and Mutagenesis in
Saccharomyces cerevisiae 267
Lyra M. Griffiths, Nicole A. Doudican, Gerald S. Shadel, and Paul W. Doetsch
18. Determination of DNA Mutation Load in Human Tissues Using Denaturing
HPLC-Based Heteroduplex Analysis 287
Kok Seong Lim, Robert K. Naviaux, and Richard H. Haas
19. Rapid Identification of Unknown Heteroplasmic Mitochondrial DNA
Mutations with Mismatch-Specific Surveyor Nuclease 301
Sylvie Bannwarth, Vincent Procaccio, and Véronique Paquis-Flucklinger
20. Collection of Isolated Cells for Studying Mitochondrial DNA Mutations
Within Individual Cells 315
*Yevgenya Kraytsberg, Natalya Bodyak, Susan Myerow, Alexander Nicholas,
Konstantin Ebrallidze, and Konstantin Khrapko*
21. Quantitative Analysis of Somatic Mitochondrial DNA Mutations by Single-Cell
Single-Molecule PCR 329
*Yevgenya Kraytsberg, Natalya Bodyak, Susan Myerow, Alexander Nicholas,
Konstantin Ebrallidze, and Konstantin Khrapko*
22. Measuring DNA Precursor Pools in Mitochondria 371
Christopher K. Mathews and Linda J. Wheeler
23. Establishment of Human Cell Lines Lacking Mitochondrial DNA 383
Kazunari Hashiguchi and Qiu-Mei Zhang-Akiyama

- Index* 393