

## **Contents**

<b>Contributors.....</b>	xvii
--------------------------	------

### **Part I Keynote Lectures**

<b>Curing Blindness with Stem Cells: Hope, Reality, and Challenges.....</b>	3
Ruben Adler	

<b>Retinal Degenerations: Planning for the Future.....</b>	21
Eliot L. Berson	

### **Part II Neuroprotection**

<b>Neurotrophins Induce Neuroprotective Signaling in the Retinal Pigment Epithelial Cell by Activating the Synthesis of the Anti-inflammatory and Anti-apoptotic Neuroprotectin D1.....</b>	39
Nicolas G. Bazan	

<b>On the Role of CNTF as a Potential Therapy for Retinal Degeneration: Dr. Jekyll or Mr. Hyde?.....</b>	45
William A. Beltran*	

<b>Nanoceria Particles Prevent ROI-Induced Blindness.....</b>	53
Junping Chen*, Swanand Patil, Sudipta Seal, and James F. McGinnis	

<b>An in-vivo Assay to Identify Compounds Protective Against Light Induced Apoptosis.....</b>	61
Yogita Kanan*, Anne Kasus-Jacobi, Kjell Sawyer, David S. Mannel, Joyce Tombran Tink, and Mauyyad R. Al-Ubaidi	

<b>Role of BCL-XL in Photoreceptor Survival.....</b>	69
Yun-Zheng Le*, Lixin Zheng, Yuwei Le, Edmund B. Rucker III, and Robert E. Anderson	

<b>The Hypoxic Transcriptome of the Retina: Identification of Factors with Potential Neuroprotective Activity.....</b>	75
Markus Thiersch*, Wolfgang Raffelsberger, Enrico Frigg, Marijana Samardzija, Patricia Blank, Olivier Poch, and Christian Grimm	

### **Part III Gene Therapy and Neuroprotection**

<b>Lentiviral Gene Transfer-Mediated Cone Vision Restoration in RPE65 Knockout Mice.....</b>	89
Alexis-Pierre Bemelmans, Corine Kostic, Maité Cachafeiro, Sylvain V. Crippa, Dana Wanner, Meriem Tekaya, Andreas Wenzel, and Yvan Arsenijevic	
<b>In vitro Analysis of Ribozyme-mediated Knockdown of an ADRP Associated Rhodopsin Mutation.....</b>	97
Dibyendu Chakraborty*, Patrick Whalen, Alfred S. Lewin, and Muna I. Naash	
<b>Gene Therapy for Mouse Models of ADRP.....</b>	107
Marina S. Gorbatyuk*, William W. Hauswirth, and Alfred S. Lewin	
<b>Development of Viral Vectors with Optimal Transgene Expression for Ocular Gene Therapies.....</b>	113
Takao Hashimoto*	
<b>Adeno-Associated Viral Vectors and the Retina.....</b>	121
John J. Alexander*, and William W. Hauswirth	
<b>Genetic Supplementation of RDS Alleviates a Loss-of-function Phenotype in C214S Model of Retinitis Pigmentosa.....</b>	129
May Nour, Steven J. Fliesler, and Muna I. Naash	
<b>Morphological Aspects Related to Long-term Functional Improvement of the Retina in the 4 Years Following rAAV-mediated Gene Transfer in the RPE65 Null Mutation Dog.....</b>	139
Kristina Narfström, Mathias Seeliger, Chooi-May Lai, Vaegan, Martin Kratz, Elizabeth P. Rakoczy, and Charlotte Remé	
<b>Virus-mediated Gene Delivery to Neuronal Progenitors.....</b>	147
Tonia S. Rex*	

### **Part IV Animal Models of Retinal Degeneration**

<b>Loss of Visual and Retinal Function in Light-stressed Mice.....</b>	157
Drew Everhart, Ana Stachowiak, Yumiko Umino*, and Robert Barlow	

<b>ERG Responses and Microarray Analysis of Gene Expression in a Multifactorial Murine Model of Age-Related Retinal Degeneration.....</b>	155
Goldis Malek, Jeffery A. Jamison, Brian Mace, Patrick Sullivan, and Catherine Bowes Rickman	
<b>Oxygen Supply and Retinal Function: Insights from a Transgenic Animal Model.....</b>	171
Edda Fahl, Max Gassmann, Christian Grimm, and Mathias W. Seeliger	
<b>Characterization of Gene Expression Profiles of Normal Canine Retina and Brain Using a Retinal cDNA Microarray.....</b>	179
Gerardo L. Paez*, Barbara Zangerl, Kimberly Sellers, Gregory M. Acland, and Gustavo D. Aguirre	
<b>Toward a Higher Fidelity Model of AMD.....</b>	185
Brian J. Raisler*, Miho Nozaki, Judit Baffi, William W. Hauswirth, and Jayakrishna Ambati	
<b>The Potential of Ambient Light Restriction to Restore Function to the Degenerating P23H-3 Rat Retina.....</b>	193
Krisztina Valter, Diana K. Kirk, and Jonathan Stone	

#### Part V Molecular Genetics and Candidate Genes

<b>Mutations in Known Genes Account for 58% of Autosomal Dominant Retinitis Pigmentosa (adRP).....</b>	203
Stephen P. Daiger, Lori S. Sullivan, Anisa I. Gire, David G. Birch, John R. Heckenlively, and Sara J. Bowne	
<b>Genetics of Age-related Macular Degeneration.....</b>	211
Albert S. Edwards	
<b>Retinal Phenotype of an X-Linked Pseudo-usher Syndrome in Association with the G173R Mutation in the <i>RPGR</i> Gene.....</b>	221
Alessandro Iannaccone, Mohammad I. Othman, April D. Cantrell, Barbara J. Jennings, Kari Branham, and Anand Swaroop	
<b>Mutation in the PYK2-Binding Domain of PITPNM3 Causes Autosomal Dominant Cone Dystrophy (CORD5) in Two Swedish Families.....</b>	229
Linda Köhn*, Konstantin Kadzhaev, Marie S.I. Burstedt, Susann Haraldsson, Ola Sandgren, and Irina Golovleva	
<b>Identification and Characterization of Genes Expressed in Cone Photoreceptors.....</b>	235
Mehrnoosh Saghizadeh*, Novrouz B. Akhmedov, and Debora B. Farber	

**Clinical and Genetic Characterization of a Chinese Family with CSNB1...245**  
Ruifang Sui\*, Fengrong Li, Jialiang Zhao, and Ruxin Jiang

<b>10q26 Is Associated with Increased Risk of Age-Related Macular Degeneration in the Utah Population.....</b>	253
D. Joshua Cameron*, Zhenglin Yang, Zhongzhong Tong, Yu Zhao, Alissa Praggastis, Eric Brinton, Jennifer Harmon, Yali Chen, Erik Pearson, Paul S. Bernstein, Gregory Brinton, Xi Li, Adam Jorgensen, Sara Schneider, Daniel Gibbs, Haoyu Chen, Changguan Wang, Kimberly Howes, Nicola J. Camp, and Kang Zhang	

**Part VI Diagnostic, Clinical, Cytopathological and Physiologic Aspects of Retinal Degeneration**

<b>Carboxyethylpyrrole Adducts, Age-related Macular Degeneration and Neovascularization.....</b>	261
--	-----

Kutralanathan Renganathan, Quteba Ebrahem, Amit Vasanji, Xiaorong Gu, Liang Lu, Jonathan Sears, Robert G. Salomon, Bela Anand-Apte, and John W. Crabb	
---	--

<b>A Possible Impaired Signaling Mechanism in Human Retinal Pigment Epithelial Cells from Patients with Macular Degeneration.....</b>	269
Piyush C. Kothary and Monte A. Del Monte	

<b>Expression and Cell Compartmentalization of EFEMP1, a Protein Associated with <i>Malattia Leventinese</i>.....</b>	277
Adam Kundzewicz*, Francis Munier, and Jean-Marc Matter	

<b>Role of ELOVL4 in Fatty Acid Metabolism.....</b>	283
Vidyullatha Vasireddy*, Majchrzak Sharon, Norman Salem, Jr., and Radha Ayyagari	

<b>Organization and Molecular Interactions of Retinoschisin in Photoreceptors.....</b>	291
Camasamudram Vijayasarathy, Yuichiro Takada, Yong Zeng, Ronald A. Bush, and Paul A. Sieving	

**Part VII Basic Science Underlying Retinal Degeneration**

<b>Proteomics Profiling of the Cone Photoreceptor Cell Line, 661W.....</b>	301
Muayyad R. Al-Ubaidi, Hiroyuki Matsumoto, Sadamu Kurono, and Anil Singh	

<b>γ-Secretase Regulates VEGFR-1 Signalling in Vascular Endothelium and RPE.....</b>	313
Michael E. Boulton, Jun Cai, Maria B. Grant, and Yadan Zhang	
<b>Analysis of the Rate of Disk Membrane Digestion by Cultured RPE Cells.....</b>	321
Tanja Diemer*, Daniel Gibbs, and David S. Williams	
<b>Functional Expression of Cone Cyclic Nucleotide-Gated Channel in Cone Photoreceptor-Derived 661W Cells.....</b>	327
J. Browning Fitzgerald, Anna P. Malykhina, Muayyad R. Al-Ubaidi, and Xi-Qin Dong	
<b>Phosphorylation of Caveolin-1 in Bovine Rod Outer Segments in vitro by an Endogenous Tyrosine Kinase.....</b>	335
Michael H. Elliott*, and Abboud J. Ghalayini	
<b>Regulation of Nuerotrophin Expression and Activity in the Retina.....</b>	343
Abigail S. Hackam*	
<b>Involvement of Guanylate Cyclases in Transport of Photoreceptor Peripheral Membrane Proteins.....</b>	351
Sukanya Karan*, Jeanne M. Frederick, and Wolfgang Baehr	
<b>Rod Progenitor Cells in the Mature Zebrafish Retina.....</b>	361
Ann C. Morris*, Tamera Scholz, and James M. Fadool	
<b>αvβ5 Integrin Receptors at the Apical Surface of the RPE: One Receptor, Two Functions.....</b>	369
Emeline F. Nandrot, Yongen Chang, and Silvia C. Finnemann	
<b>Implantation of Mouse Eyes with a Subretinal Microphotodiode Array.....</b>	377
Machelle T. Pardue, Tiffany A. Walker, Amanda E. Faulkner, Moon K. Kim, Christopher M. Bonner, and George Y. McLean	
<b>Variation inthe Electoretinogram of C57BL/6 Substrains of Mouse.....</b>	383
Alison L. Reynolds*, G. Jane Farrar, Pete Hu mphries, and Paul F. Kenna	
<b>A2E, A Pigment of RPE Lipofuscin, is Generated from the Precursor, A2PE by a Lysosomal Enzyme Activity.....</b>	393
Janet R. Sparrow, So Ra Kim, Ana M. Cuervo and Urmila Bandhyopadhyay and	
<b>Endothelin Receptors: Do They Have a Role in Retinal Degeneration?.....</b>	399
Vanessa Toribidoni*, María Iribarne, and Angela M. Suburo	

<b>CNTF Negatively Regulates the Phototransduction Machinery in Rod Photoreceptors: Implications for Light-Induced Photostasis Plasticity.....</b>	407
Rong Wen, Ying Song, Yun Liu, Yiwen Li, Lian Zhao, and Alan M. Laties	
<b>About the Editors.....</b>	415
<b>Index.....</b>	419