CURRICULUM VITAE

Paul Albert Sieving, M.D., Ph.D.
Director, National Eye Institute, National Institutes of Health
Member, Institute of Medicine, The National Academies

EDUCATION

Valparaiso University, 1966–1970, Physics and History, B.A. with Highest Distinction
Yale University Graduate School, 1970–1973, Physics, M.S.
Yale Law School, 1973–1974; J.D. degree program; leave of absence, 1974
University of Illinois Medical School, 1974–1978, M.D.
University of Illinois Graduate School, 1976–1981, Biomedical Engineering, Ph.D.

POSTDOCTORAL AND FELLOWSHIP TRAINING

Medical Intern: University of Illinois Hospital, 1978–1979
Residency in Ophthalmology, University of Illinois Eye and Ear Infirmary, Chicago, 1979–1982
Postdoctoral Fellowship in Retinal Physiology: Roy H. Steinberg, M.D., Ph.D., Professor of Physiology, University of California, San Francisco, 1982–1984
Medical Fellowship in Inherited Retinal Degenerations: Eliot L. Berson, M.D., Professor of Ophthalmology, Berman-Gund Laboratory for Retinal Degenerations, Harvard Medical School, 1984–1985

ACADEMIC APPOINTMENTS

Assistant in Ophthalmology, Illinois Eye & Ear Infirmary, University of Illinois, Chicago, 1979–1982
Visiting Assistant Research Ophthalmologist, University of California, San Francisco, 1982–1984
Fellow in Ophthalmology, Harvard Medical School, Massachusetts Eye & Ear Infirmary, 1984–1985
University of Michigan 1985–2001:
- Department of Ophthalmology, Asst. Prof., 1985–89; Assoc. Prof., 1989–94; Professor, 1994–2001
- Paul R. Lichter Professor of Ophthalmic Genetics, 1990–2001 (leave of absence, 2001–current)
- Rackham Graduate School, Faculty: Neuroscience Program 1985–2001; Bioengineering, 1985–2001
- Founding Director, Center for Retinal and Macular Degenerations, 1990–2001
- Center for Biomedical Engineering Research (CBER), 1997–2001
- Founding Director, Ophthalmic Molecular Diagnostics Laboratory, CLIA, Certified, 1999–2001

CURRENT POSITIONS

Director, National Eye Institute, National Institutes of Health; Bethesda, Maryland; 2001–current
Tenured Senior Investigator, NIH Intramural Research Program; Bethesda, Maryland; 2001–current

LICENSURE

National Board of Medical Examiners, Diplomat, 1978
American Board of Ophthalmology, Diplomat, 1983
Medical Licenses: Illinois, 1978; California, 1982; Massachusetts, 1984; Michigan, 1985

AWARDS AND HONORS

Graduated "With Highest Distinction," Valparaiso University, 1970
Yale University: Graduate School Fellowship, 1970–1973; Law School Fellowship, 1973
University of Illinois Medical School: James Scholar, 1974–78; Leon F. Moldavsky Physiology Award, 1977; College of Medicine Research Award, 1978; Sigma Xi, 1980
Fight-for-Sight Research Grant Award, 1980–1982
Career Development Award, National Retinitis Pigmentosa Foundation, 1982–1985
Olga Keith Wiess Scholar, Research to Prevent Blindness, Inc., 1989
Distinguished Alumnus, Valparaiso University, 1991
American Ophthalmological Society, Member, 1993 (thesis required for membership)
Research to Prevent Blindness Senior Scientific Investigator Award, 1998
Alcon Award, Alcon Research Institute, 2000 (5 awards/year worldwide)
The Best Doctors in America, 1996–1997 (Midwest); 1998, 2001, 2005
Who's Who in America, 2003–2009; Who's Who in Medicine and Healthcare, 2002–2007; Who's Who in Science and Engineering, 2005–2007; Who's Who in the East, 2006–2008; Who's Who in American Politics, 2009–2010
Academia Ophthalmologica Internationalis, elected 2004 (60 elected "chairs" worldwide)
Pisart Vision Award, Lighthouse International for the Blind, New York, 2005 (1 award/year worldwide)
The Retina Society, Associate Member, elected 2005
American Academy of Ophthalmology, Achievement Award, 2005
Institute of Medicine of the National Academies, elected 2006
Health Care Leadership Award, American Optometric Association, 2007
National Institutes of Health, Office of Equal Opportunity and Diversity Management, Making a Difference Award, 2009

DEPARTMENTAL AND COMMITTEE SERVICE (University Michigan, through 2001)

MSTP Program Mentor, University of Michigan, 1985–2001
Preparation of Ophthalmology Department NEI/NIH CORE Grant for Vision (successful funding), 1985
Director, Vivarium Module, NIH/NEI Vision Core Grant, 1986–1992
Chair, Faculty Recruitment and Promotions Committee, Ophthalmology Department, 1987–2001
Ophthalmology Department Internal Review Committee, Dean's Periodic Review, 1987
Executive Committee, Ophthalmology Department, 1989–1992
Director, NIH Vision Training Grant, University of Michigan, 1989–1992
Examiner, UM Medical School Comprehensive Clinical Assessment, 1992–1993
Speaker, University of Michigan Medical School Alumni Meeting, Phoenix, AZ, 3/1993
Ophthalmology Department Internal Review Committee, Dean's Periodic Review, 1994
Faculty search committee: Dept. of Biomedical Engineering position in applied laser engineering, 1997
Searle Selection Committee, UM Medical School, 1999
Burroughs Wellcome Fund Clinical Scientist Awards selection committee, UM Medical School, 1999
Advisory Council on Clinical Research (ACCR), UM Medical School, 1997–2000

NIH LEADERSHIP, COMMITTEES AND OTHER SERVICE

Electronic Research Administration (eRA) oversight committee, Chair, 2002–2003
NIH Central Compensation Committee, Member, 2002–Nov. 2004
Search committee for Director of NIMH, Member, 2002
Search committee for Director of Center for Scientific Review (CSR), Co-chair, 2003–2004

Roadmap Structural Biology Working Group, 2003–continuing
Nanotechnology/Nanomedicine Working Group, Co-chair, 2003
NanoMedicine Roadmap Initiative, Co-chair, 2003–continuing
RICC Roadmap Implementation Coordination Committee, 2003–continuing
Search committee for director of NIEHS, Member, Spring, 2004
Neuroscience Blueprint: NIH Directors' Development Committee, 2004–continuing
NIH Management and Budget Working Group, Member, 11/2004–5/2006
NIH Intramural Working Group, Co-chair, 5/2006–continuing
NIH Steering Committee, 2006–continuing

ADVISORY BOARDS

NIH/NEI Review and Study Sections:
Visual Sciences C, 1990–1993; Visual Sciences A2, *ad hoc* 1986–1989; Special Reviewers Reserve, 1993–1998
Program Panel: *Research Strategies for Age-Related Macular Degeneration*, 1988
Co-chair, Retinal Diseases Section, *Vision Research: A National Plan*, 1999–2003
VA Research Study Section (Vision), Washington, D.C., November 1987
Fight-for-Sight, Grants-in-Aid Review Panel, 1988–1991
Foundation Fighting Blindness, Scientific Advisory Board, 1989–2001; Vice-chair Clinical Research, 1995–2001
Editorial Boards: *Experimental Eye Research*, 1997–2001; *Vision Research*, 1992–2000
Bressler Award Committee, Jewish Guild for the Blind, New York City, 2002–current
Champalimaud Foundation, Award Committee, Lisbon, Portugal, 2006–current

MEMBERSHIPS

Association for Research in Vision and Ophthalmology (ARVO), 1977–current
International Society for Clinical Electrophysiology of Vision, 1978–current; Treasurer, 1986–1994
American Association for the Advancement of Science, 1980–current
American Academy of Ophthalmology, 1980–current
American Society for Neuroscience, 1988–current
American Society of Human Genetics, 1989–current
American Ophthalmological Society, 1993–current
Academia Ophthalmologica Internationalis, 2004–current
The Retina Society, Associate Member, 2005–current
Institute of Medicine of the National Academies, 2006–current

GRANT AND PROGRAM SUPPORT

University of Michigan (Continuous Support as Principal Investigator, 1986–2001)
Inner Retinal Contributions to the ERG
NIH R01-EY06094, 1986–2003, 1998–2003, TDC: $986,224
PI: P.A. Sieving; transferred to Bret Hughes, 6/2001
Retinoschisis: Genetic Linkage and Positional Cloning
NIH R01-EY10259, 1993–2000, 1996–2000, TDC: $753,000
PI: P.A. Sieving
Michigan Retinal and Macular Degeneration Center

The Foundation Fighting Blindness, Hunt Valley, MD; 1988–2005, 2000–2005, TDC: $2,297,298
PI: P.A. Sieving; transferred to Anand Swaroop, 6/2001

Medical Therapy for Retinal and Macular Degenerative Disease
The Foundation Fighting Blindness, Hunt Valley, MD, 1998–2001, TDC: $625,000
PI: P.A. Sieving

RPB Senior Scientific Investigator Award, 1998 (P.A. Sieving)
Research to Prevent Blindness: $50,000

Genomic Technology and Genetic Disease, NIH Human Genome Project
Pilot Project Leader: P.A. Sieving, 1990–1995
PI: F.S. Collins

NIH 1P30 EY07003, Core Grant for Vision Research,
Module Director: P.A. Sieving, 1987–1992, TDC: $740,000
PI: P.R. Lichter

Michigan Vision Research Training Grant: NIH T32 EY07022
1989–1992, TDC: $110,000
PI: P.A. Sieving

Olga Keith Weiss Scholar, 1989, P.A. Sieving: Research to Prevent Blindness, $30,000.

Studies of Inherited Retinal Degenerations, Michigan Eye Bank and Transplantation Center
1986–1988, TDC: $20,600
PI: P.A. Sieving

Molecular Genetics of Retinal Dystrophies, The Foundation Fighting Blindness, Hunt Valley, MD
1989–1991, TDC: $300,000
PI: P.A. Sieving

Career Development Award, P.A. Sieving
National Retinitis Pigmentosa Foundation, Baltimore, MD, 1982–1985, $120,000

Principal Investigator, Section on Translational Research for Retinal and Macular Degeneration, NIH Intramural Research Program

Human Protocol NIH 03-EI-0033: “X-linked Juvenile Retinoschisis: Clinical and Molecular Studies” (PI: P.A. Sieving, 2003)

Human Protocol NIH 03-EI-0179: “Investigation of the Effect of Dietary Docosahexaenoic Acid (DHA) Supplementation on Macular Function in Subjects with Autosomal Dominant Stargardt-like and Autosomal Recessive Stargardt Macular Dystrophy” (PI: P.A. Sieving, 2003)

Human Protocol NIH 03-EI-0234: “A Phase I Study of NT-501-10 and NT-501-6A.02, Implants of Encapsulated Human NTC-210 Cells Releasing Ciliary Neurotrophic Factor (CNTF), in Patients with Retinitis Pigmentosa” (PI: P.A. Sieving, 2003)

Human Protocol NIH 03-EI-0255: “Pilot Study on the Effect of Vitamin A Supplementation on Cone Function in Retinitis Pigmentosa (RP)” (PI: P.A. Sieving, 2003)

Human Protocol 06-EI-0071: “A Phase-2 Study of Implants of Encapsulated Human NTC-201 Cells Releasing Ciliary Neurotrophic Factor (CNTF) in Participants with Visual Acuity Impairment Associated with Atrophic Macular Degeneration” (PI: P.A. Sieving, 2006)

Laboratory Section Protocol NIH Z01 DC000065-0: “Retinal Degeneration: Genetic and Environmental Animal Models” (PI: P.A. Sieving, 2002)

POST-DOCTORAL FELLOWS

Kenji Wakabayashi, M.D., 1986–1988, Asst. Professor of Ophthalmology, Kanazawa University, Japan; Subsequent position: private practice of ophthalmology, Kanazawa, Japan.

Franklin Naarendorp, Ph.D., 1987–1992;
Subsequent position: Associate Professor of Psychology, Northeastern University, Boston, MA.
Lian Shentu, Ph.D., 1988–1989;
Subsequent position: Research Scientist, Peripheral Dynamics, Inc., Plymouth Meeting, PA.
Koichiro Murayama, M.D., 1989–1991;
Subsequent position: Professor of Ophthalmology, Saitama Medical School, Saitama, Japan.
Zhengping Zhuang, M.D., Ph.D., 1992–1993, Molecular Genetics Postdoctoral Fellow;
Subsequent position: National Cancer Institute/NIH, Bethesda, MD.
Ronald Bush, Ph.D., 1991–1994, Retinal Physiology Postdoctoral Fellow;
Subsequent positions: Assistant Research Scientist, Department of Ophthalmology, University of Michigan, Ann Arbor, MI; Staff Scientist, Section on Translational Research for Retinal and Macular Degeneration, National Institute on Deafness and Other Communication Disorders/NIH, Bethesda, MD.
John Sullivan, M.D., Ph.D., 1992–1994, Clinical Research Fellow;
Subsequent position: Assistant Professor of Ophthalmology & Cell Biology, SUNY Syracuse, NY.
Hemant Pawar, Ph.D., 1993–1995. Molecular Genetics Postdoctoral Fellow;
Subsequent position: Research Investigator, Ophthalmology, Washington University School of Medicine, Saint Louis, MI.
Michael Fowler, Ph.D., 1993–1996, Molecular Genetics Postdoctoral Fellow;
Subsequent position: Research Assistant Professor, University of Rochester, NY.
Kelaginamane T. Hiriyanna, Ph.D., 1994–1996, Molecular Genetics Postdoctoral Fellow;
Subsequent position: Research Investigator, Dept. of Ophthalmology, University of Michigan, Ann Arbor, MI.
Kazushige Toda, M.D., 1996–1998, Retinal Physiology Fellow;
Subsequent position: Jikei University Medical School, Tokyo, Japan.
Yumiko Toda, M.D., 1996–1999, Fellow in Clinical Retinal Dystrophies;
Subsequent position: Jikei University Medical School, Tokyo, Japan.
Takeshi Sugawara, M.D., Ph.D., 1996–1998, Retinal Physiology Fellow;
Subsequent position: Assistant Professor of Ophthalmology, Iwate Medical School, Morioka, Japan.
Christine McHenry, Ph.D., 1996–1999, Molecular Genetics Postdoctoral Fellow;
Subsequent position: Research Associate, Ophthalmology and Visual Science, University of Michigan, Ann Arbor, MI.
Bo Lei, M.D., Ph.D., 1997–2001, Retinal Physiology Fellow;
Subsequent position: Assistant Professor of Ophthalmology, University of Missouri, Columbia, MO.
Shigeki Machida, M.D., Ph.D., 1998–2000, Retinal Physiology Fellow;
Subsequent position: Assistant Professor of Ophthalmology, Iwate Medical School, Morioka, Japan.
Mineo Kondo, M.D., Ph.D., 1999–2001, Retinal Physiology Fellow;
Subsequent position: Associate Professor of Ophthalmology, University of Nagoya Medical School, Nagoya, Japan.
Yuichiro Takada, M.D., Ph.D., 2000–2005. Retinal Physiology Fellow.
Atsuhiro Tanikawa, M.D., Ph.D., 2001–2003, Retinal Physiology Fellow;
Subsequent position: University of Nagoya Medical School, Nagoya, Japan.
Dorit Raz, D.V.M., 2003–2006, Retinal Physiology Fellow.
Subsequent position: Department of Neurobiology, Tel-Aviv University, Tel-Aviv, Israel
Sten Kjellstrom, M.D., 2003–2006, Retinal Physiology Fellow;
Subsequent position:
Masatoshi Haruta, M.D., Ph.D., 2005–2008, Retinal Physiology Fellow;
Subsequent position: Hyogo Prefectural Amagasaki Hospital, Hyogo, Japan

Tae Kwann Park, M.D., 2007–2009, Retina Physiology Fellow;
Subsequent position: Soonchunhyang University Bucheon Hospital, Bucheon, South Korea
Lucia Ziccardi, M.D., 2008–present, Retina Physiology Fellow.
Hongman Song, M.D., Ph.D., 2009–present, Retina Physiology Fellow.

GRADUATE STUDENTS

William Lemon (M.S., 1987), Bioengineering, University of Michigan, Ann Arbor, MI
Andrew Geller (Ph.D., 1992), Psychology, University of Michigan, Ann Arbor, MI
Jeffrey Jamison (Ph.D., 2003), Bioengineering, University of Michigan, Ann Arbor, MI
Joshua Fernandes (M.D.), Medicine, George Washington University School of Medicine and Health Sciences, Washington, DC

UNIVERSITY OF MICHIGAN, COURSE AND TEACHING ACTIVITIES (through 2000)

Ophthalmology 804, "Introduction to Clinical Vision Science," course director, Rackham Graduate School, 1986, 1988
Physiology 733, "Introduction to Basic Vision Science," Rackham Graduate School, 1989–1990
Neuroscience 700, organizer of student seminars in "Photoreceptor Contribution to the ERG." Rackham Graduate School, 1990
UM Ophthalmology Department Outreach Program, "Inherited Retinal Dystrophies," Grand Rapids, MI, November, 1990
UM Ophthalmology Department CME Course: Interpreting Fluorescein Angiograms in Retinal Vascular Diseases, "Central Retinal Vein Occlusion," 4/1991
American Academy of Ophthalmology, Annual National Meeting, "Introduction to Ophthalmic Molecular Genetics," Course Organizer and Director, 1989–1991
UM Ophthalmology Spring Postgraduate Course: Lasers in Ophthalmology, Applications and New Developments, "The Role of Laser Therapy in Central Retinal Vein Occlusion," 5/1991
UM Ophthalmology Department Outreach Program, "DNA Gene Mutations in Retinitis Pigmentosa," Midland Hospital, Midland, MI, 2/1992
Kresge Eye Institute, Wayne State University, Clinical and Basic Lecture Series in Ophthalmology: "Retinal Physiology of Inherited Retinal Dystrophies," Detroit, MI, 3/1992
UM Ophthalmology Department CME Course: Retina Update, "From Laboratory to Clinic: Impact of Molecular Diagnosis," 3/1993
University of Michigan School of Public Health, Toxicology Seminars 601, "Toxicology and Retinal Disease," 11/1994
UM Ophthalmology Department. Retina Update CME Course, "Molecular Genetics for the Ophthalmologist," 4/1995
UM Ophthalmology Department, Retina CME Course, "Inherited Retinal Dystrophies/Vitamin A Therapy," 4/1996
UM Ophthalmology Department, "1996 Harold F. Falls Symposium: The X-chromosome and Retinal Disease," Symposium organizer with 13 invited speakers, Ann Arbor, MI, 6/1996
University of Michigan Ophthalmology Department, Retina CME Course, "Molecular Diagnosis in the 21st Century," 6/1997
UM Medical Center Health Night Out, "Macular Degeneration," Ann Arbor, MI, 10/1997
UM Ophthalmology Residents, "Color Vision," 1/1998
UM Ophthalmology Residents, "Hereditary Retinal Degenerations," 1/1998
Henry Ford Hospital Resident Lecture Series, "Retinal Dystrophies and Physiology," 3/1998

UM Ophthalmology Department Outreach Series, "Retinal and Macular Degeneration: New Genes and More Genes," Bay City, MI, 3/1998
University of Chicago, Ophthalmology Department, "Transgenic Mouse Models of Retinal Degeneration," 3/1998
Kresge Eye Institute, Wayne State University, Visiting Professor: "Genotypes, Phenotypes, and Physiology of Hereditary Retinal and Macular Dystrophies," Detroit, MI, 3/1998
Bioengineering 496 Lecture, "Bioengineering approaches to retinal signaling," Rackham Graduate School, 1990, 1993, 1998
Supervising Faculty, Bioengineering 599, "Graduate Student Independent Research," Rackham Graduate School, 1988–9, 1992, 1994–9
UM Ophthalmology Residents, "Electrophysiology and Visual Psychophysics," 1/1999
"Neuroscience Student Research," Rackham Graduate School, 1999
Supervising Faculty, Bioengineering 990, "Graduate Student Research," Rackham Graduate School, 1999
Kresge Eye Institute, Wayne State University, Clinical and Basic Lecture Series in Ophthalmology, "ERG, Genetics and Clinical RP," Detroit, MI, 3/2000
Henry Ford Ophthalmology Visiting Professor, "Retinal and Macular Dystrophies," Detroit, MI, 4/2000

CLINICAL RESEARCH ACTIVITIES (NIH)

Human Protocol NIH 03-EI-0033: "X-Linked Juvenile Retinoschisis - Clinical and Molecular Studies." (PI: Sieving PA, 2003–current)
Human Protocol NIH 03-EI-0179: "Investigation of the Effect of Dietary Docosahexaenoic Acid (DHA) Supplementation on Macular Function in Subjects with Autosomal Dominant Stargardt-Like and Autosomal Recessive Stargardt Macular Dystrophy." (PI: Sieving, 2003–2007)
Human Protocol NIH 03-EI-0234: "A Phase I Study of NT-501-10 and NT-501-6A.02, Implants of Encapsulated Human NTC-210 Cells Releasing Ciliary Neurotrophic Factor (CNTF), in Patients with Retinitis Pigmentosa." (PI: Sieving PA, 2003–2006)
Human Protocol NIH 03-EI-0255: "Pilot Study on the Effect of Vitamin A Supplementation on Cone Function in Retinitis Pigmentosa (RP)." (PI: Sieving PA, 2003–2009)
Human Protocol NIH 06-EI-0071: "A Phase 2 Study of Implants of Encapsulated Human NTC-201 Cells Releasing Ciliary Neurotrophic Factor (CNTF), in Participants with Visual Acuity Impairment Associated with Atrophic Macular Degeneration." (PI: Sieving PA, 2006–)
Laboratory Section Protocol NIH Z01 DC000065-0: "Retinal Degeneration: Genetic and Environmental Animal Models." (PI: Sieving PA, 2002–current)

INVITED SEMINARS AND LECTURES

1987–2001

Oakland University Eye Institute, "Electrophysiological Studies of Human Retinal Dystrophies," Rochester, MI, 10/1987
Japanese Ophthalmological Society, Annual Meeting, Keynote speaker, "Electrophysiology of the Inner Retina: Diagnosis Using New ERG Components," Kyoto, Japan, 3/1988
Kanazawa University Department of Ophthalmology, "Molecular Genetics of Juvenile Retinoschisis," Kanazawa, Japan, 3/1988
Kaio University Vision Seminars, "Application of Molecular Genetics to Human Retinal Degeneration," Tokyo, Japan, 3/1988
International Society for Clinical Electrophysiology of Vision (ISCEV), Annual Meeting, Keynote Lecture, "The Scotopic Threshold Response of the Electroretinogram," Lisbon, Portugal, 6/1988

Illinois Eye and Ear Infirmary, "Contributions to the Electroretinogram from the Inner Retina of the Cat," Chicago, IL, 4/1989
Kresge Eye Institute, Wayne State University, "Molecular Genetics of Retinal Dystrophies," Detroit, MI, 12/1989
American Academy of Optometry, Annual Meeting, Symposium on Physiological Optics, Invited Speaker, "Analysis of the Rod ERG," New Orleans, LA, 12/1989
Color Vision Symposium Festschrift for Mathew Alpern, University of Michigan, Symposium Invited Speaker, "Scotopic Threshold Response: Monitoring Rod Vision in Starlight by the ERG," 5/1990
St. Joseph–Mercy Hospital Ophthalmology Meeting, "Molecular Genetics of Retinal Dystrophies," Ypsilanti, MI, 7/1991
Michigan State University, Clinical Electrophysiology Symposium, Symposium Invited Speaker, "On- and Off-pathway Activity in the Primate ERG," East Lansing, MI, 10/1991
Detroit Ophthalmological Society, "Tribute to Harold Falls: Ophthalmic Molecular Genetics," 10/1991
Kresge Eye Institute, Wayne State University, Clinical and Basic Lecture in Ophthalmology, "Clinical Physiology of Inherited Retinal Dystrophies," Detroit, MI, 3/1992
Temple University, Ophthalmology Department Grand Rounds, "Molecular Genetics of Inherited Macular Dystrophies," Philadelphia, PA, 12/1992
University of Utah, Ophthalmology Department, "Molecular Genetics of Inherited Retinal Dystrophies," Salt Lake City, UT, 4/1993
FASEB, The Biology and Chemistry of Vision, Session Organizer and Speaker, "Causes of Human Retinal Degeneration," Copper Mountain, CO, 6/1993
Swiss Ophthalmic Research Club, Annual Meeting, Keynote Speaker, "Ophthalmic Molecular Genetics: Techniques, Applications, and Status," Leysin, Switzerland, 3/1994
Ophthalmic Club of Toledo, "Molecular Genetics for the Ophthalmologist," Toledo, OH, 3/1995
University of West Virginia, Dept. of Ophthalmology, 16th Annual Conference, Keynote Speaker, "Genotypes, Phenotypes, and Physiology: Ophthalmic Molecular Genetics." Morgantown, WV, 10/1995
Macula Workshop, "What Limits Vision in Photoreceptor Dystrophies Besides Quantum Catch Loss?," Bath, England, 6/1996
International Society for Clinical Electrophysiology of Vision, Keynote Speaker, "Primate Photopic ERG: New Concepts of Response Origins," Tübingen, Germany, 7/1996
VII International Symposium on Retinal Degeneration, Invited Speaker, "Reinterpreting the Origins of Photopic ERG Components," Sendai, Japan, 10/1996
University of Alabama, Loris and David Rich Lecture Series in Visual Sciences, "Retinal Neural Signaling of On- and Off-pathway Cells," Birmingham, AL, 11/1996
The Association for Research in Vision and Ophthalmology 1997, Invited Speaker, Retinitis Pigmentosa Interest Group, "How Many Photoreceptors Do We Need for Vision?," Ft. Lauderdale, FL, 5/1997
Fernstrom Symposium 1997, Symposium Speaker, "Mouse Model of G90D Rhodopsin Night Blindness," Lund, Sweden, 6/1997
Third Great Basin Visual Science Symposium. Symposium Speaker, "Night Blindness in Man and Mouse: The G90D Rhodopsin Mutation," Salt Lake City, UT, 8/1997
OSA Annual Meeting and ILS-XIII, "The A-wave at Scotopic and Photopic Threshold: Rethinking the Origins of the ERG," Long Beach, CA, 8/1997
The University of Illinois at Chicago, Department of Ophthalmology and Visual Sciences and the UIC Eye Center Alumni Association, 1998 Peter C. Kronfeld Memorial Lecturer, "How Many Photoreceptors Do We Need for Vision?" 6/1998
Belgium Ophthalmological Society, "Genetics of Retinal Dystrophies," Brussels, Belgium, 11/1998

The Association for Research in Vision and Ophthalmology, Organizer and Speaker, "ARVO Basic and Clinical Science Lecture: Photoreceptors from a Clinical Point of View," 5/9/1999

The Association for Research in Vision and Ophthalmology, Session Moderator, "Mutations Associated with Macular Disease," 5/13/1999

FASEB, The Biology and Chemistry of Vision, "Dark Adapted Visual Function and Human Retinal Disease," Session Organizer and Moderator, "Photoreceptor Disease and Visual Thresholds," Copper Mountain, CO, 6/13–18/1999

Institute of Ophthalmology, University College, London, "P23H Rhodopsin Mutation: Human RP and Transgenic Animal Model," London, England, 10/5/1999

University of Rochester, Neuroscience Seminar Series, "Retinitis Pigmentosa: Human Disease and Animal Models," Rochester, NY, 11/1999

Royal College of Ophthalmologists, Annual Meeting, "American Experience with Ophthalmic Diagnostic Genetics Service," Invited Lecture, Harrogate, England, 5/24/2000

European Concerted Action Meeting, Invited Speaker, "Therapy for Photoreceptor Disease by Manipulation of RPE Retinoid Pathway," Sintra, Portugal, 7/2000

Scandinavian 30th Annual Ophthalmology and Vision Research Meeting, Plenary Lecturer, "Animal Models of Retinal Degenerations" and "Diagnostic Molecular Genetics for Human Retinal Degenerations," Karlskrona, Sweden, 9/1–3/2000

ISCEV–Japan, Annual Meeting, Keynote Speaker, "Electrophysiology of Animal Models of Retinal Degeneration," Morioka, Japan, 9/2000

University College, Institute of Ophthalmology, Vision Research Symposium, Davson Lecture, "Nightblindness and the State of Rod Cells in Retinitis Pigmentosa," London, England, 11/29/2001

American Academy of Optometry Annual Meeting, Keynote Speaker, Philadelphia, PA, 12/6/2001

2002

Columbia University, Department of Ophthalmology, Smeltzer Lecture, "Nightblindness and the State of Rod Cells in Rhodopsin Mutations," New York, NY, 1/10/2002

American Ophthalmological Society, Verhoeff Lecture, Sea Island, GA, 5/19/2002

Howard University, Department of Ophthalmology, Lois A. Young Memorial Lecture, "Ophthalmic Molecular Diagnostic Testing," Washington DC, 6/7/2002

Louisiana State University, Chancellor's Award Lecture in Neuroscience and Ophthalmology, "Two Physiological Types of Nightblindness from Rhodopsin Mutations," New Orleans, LA, 11/20/2002

2003

Schepens Eye Research Institute, Harvard University, Broadhurst Distinguished Lecture, "Nightblindness and the State of Rod Cells in Retinitis Pigmentosa," Boston, MA, 2/10/2003

University of Michigan, Department of Ophthalmology and Visual Sciences, The Harold F. Falls Chair Inauguration Lecture, 5/30/2003

Duke University, Department of Ophthalmology, Bryan Research Lecturer, "Searching for Clues to AMD in Monogenetic Macular Degenerations," Durham, NC, 10/9/2003

Washington Advisory Group workshop on "Emerging Therapies for Visual Impairment," Invited Speaker, San Diego, CA, 10/14/2003

Association for Research in Vision and Ophthalmology (ARVO) Board of Trustees, Invited Speaker, Rockville, MD, 10/24/2003

Macular Vision Research Foundation, Invited Speaker, Philadelphia, PA, 10/25/2003

American Academy of Ophthalmology (AAO) Annual National Meeting, Retina Sub-specialty Day, Invited Speaker, "New Thoughts on Juvenile Retinoschisis," Anaheim, CA, 11/15/2003

2004

Association of University Professors of Ophthalmology (AUPO) Clinician Scientist Symposium, Speaker, Sarasota, FL, 1/28/2004

Foundation Fighting Blindness, "For the Love of Sight," Invited Speaker, Washington DC, 2/11/2004
University of Maryland. Neuroscience Program, Invited Lecturer, Baltimore, MD, 2/12/2004
National Eye Health Education Program (NEHEP) Conference Plenary Speaker, Charleston, SC, 3/1/2004
SUNY Syracuse. Neuroscience Program, Syracuse, NY, 3/3/2004
Columbia University, Department of Ophthalmology. Age-related Macular Degeneration Symposium on Pathogenesis and Therapeutic Strategies, New York, NY, 3/19/2004
American Association for Pediatric Ophthalmology and Strabismus (AAPOS) Annual Meeting, Plenary Speaker, Washington DC, 3/30/2004
Japanese Ophthalmological Society Meeting, Plenary Speaker, "X-linked Retinoschisis: From Gene to Treatment," Tokyo, Japan, 4/18/2004
The Association for Research in Vision and Ophthalmology 2004, The Scientific and Medical Advisory Board of Retina International, Invited Speaker, Ft. Lauderdale, FL, April, 2004
West Virginia University Center for Neuroscience, Dedication and Symposium, Keynote Speaker, Morgantown, WV, 6/2/2004
XIth International Symposium on Retinal Degeneration, Ian Constable Lecture, "X-linked Retinoschisis as a Case Study in the Evolution of Disease Understanding," Perth, Australia, 8/23/2004
Jackson Laboratory, "Laboratory Mouse in Vision Research Meeting," Keynote Speaker, "Supporting Ocular Genetic Research: Mice and Other Issues," Bar Harbor, ME, 10/15/2004
American Academy of Ophthalmology (AAO) Annual National Meeting, Retina Sub-specialty Day, Invited Speaker, "Ophthalmic Diagnostic Genotyping," New Orleans, LA, 10/23/2004
Academia Ophthalmologica Internationalis Oration, "Juvenile X-linked Retinoschisis and the Evolution of Medical Knowledge," New Orleans, LA, 10/23/2004
Society for Neuroscience, First Public Announcement of the NIH New "Neuroscience Blueprint," San Diego, CA, 10/25/2004
American Society for Human Genetics, Symposium Moderator, "The Ophthalmologists' View of Genetics," Toronto, Canada, 10/27/2004
National Neurovision Research Institute, "The NEI Translational Clinical Research Initiatives on Inherited and Orphan Retinal Diseases," Washington DC, 11/5/2004
International Society for Clinical Electrophysiology of Vision (ISCEV), Keynote Speaker, "Origins of ERG Responses in the Primate Retina," San Juan, PR, 11/15/2004
NIDCD Research Retreat, Report of the Laboratory Section of Translational Research in Retinal and Macular Degeneration, Airlie House, VA, 11/18/2004
Dean McGee Eye Institute, University of Oklahoma, New Building Announcement, Oklahoma City, OK, 11/22/2004
David G. Cogan Clinician Scientist Symposium, Speaker, NEI/NIH, Bethesda, MD, 12/13/2004

2005

Doheny Eye Insitute, University of Southern California, Presentation to Residents, "X-linked Retinoschisis: A Case Study in the Evolution of Disease Concepts," Los Angeles, CA, 1/10/2005
University of California, San Francisco, Roy H. Steinberg Memorial Lecture, San Francisco, CA, 1/13/2005
Smith-Kettlewell Eye Research Institute, "Research Opportunities: Ocular Genetics, Neuroscience, and Nanomedicine," San Francisco, CA, 1/14/2005
William Beaumont Hospital, Ropard Foundation Symposium, Keynote Speaker, "Juvenile X-linked Retinoschisis," Royal Oak, MI, 5/7/2005
Academia Ophthalmologica Internationalis, "Ophthalmic Genetic Disease—Medicine in the 21st Century," Florence, Italy, 5/11/2005

American Academy of Ophthalmology (AAO) Annual Meeting, Retina Sub-specialty Day, Invited Speaker, "A Safety Study of CNTF Implants for Human Retinitis Pigmentosa Therapy," Chicago, IL, 10/15/2005

University of Wisconsin, Department of Ophthalmology and Visual Sciences, Day-long educational symposium for 700 individuals with age-related macular degeneration, "Macular Degeneration—Progress in Sight V," Invited Speaker, "Nutritional Intervention, Pharmacologic Rescue Factors and Genetic Studies in Macular Degeneration," Madison, WI, 10/11/2005

Lighthouse International, Mrs. Cecilia Schupf and Paul J. Schupf Annual Symposium, Award Lecture, "From Concepts to Treatments: Diabetic Retinopathy, Neuroscience, and Genetics Opportunities," New York, NY, 10/21/2005

Tübingen University Eye Hospital, Symposium on the Function and Dysfunction of Vision, Invited Lecture, "Human Retinal Neurodegeneration Therapy with CNTF: Phase-1 Study," Tübingen, Germany, 11/4/2005

2006

World Ophthalmology Congress, Invited Speaker, "Genetics and the Future of Ophthalmology," São Paulo, Brazil, 2/21/2006

Washington University, St. Louis Ophthalmological Society, Invited Speaker, "Vision Research and the National Eye Institute," 3/16/2006

The Association for Research in Vision and Ophthalmology 2006, Plenary Keynote Speaker, "Vision Research: Building Productive Partnerships Across Geographic Boundaries," Ft. Lauderdale, FL, 4/30/2006

The Association for Research in Vision and Ophthalmology 2006, Invited Speaker, "The Promise of Nanotechnology," Ft. Lauderdale, FL, 5/1/2006

Federation of American Societies for Experimental Biology (FASEB), "Juvenile X-linked Retinoschisis, Electronegative ERGs, and the Search for Synaptic Dysfunction," Indian Wells, CA, 7/19/2006

John A. Moran Eye Center, 2006 Moran Symposium on Retinal Degeneration, Invited Lecture, "Treatment of Retinal Dystrophies," Salt Lake City, UT, 8/2/2006

Chinese Ophthalmological Society, Third Global Chinese Ophthalmic Conference, "Nanomedicine in Ophthalmology" and "Ciliary Neurotrophic Factor (CNTF) Phase-1 Clinical Trial for Retinal and Macular Degeneration," Beijing, China, 9/1/2006

European Association for Vision and Eye Research (EVER), ACTA Keynote Lecture, "The Interplay of Clinical and Basic Knowledge in X-linked Retinoschisis," Vilamoura, Portugal, 10/5/2006

European Paediatric Ophthalmological Society (EPOS), Keynote Lecture, "Ciliary Neurotrophic Factor (CNTF): Background and Outcome of a Phase 1–2 Clinical Trial for Retinal Degeneration," Vilamoura, Portugal. 10/6/2006

HHMI-NIH Research Scholars and the NIH Clinical Research Training Program Fellows Seminar, Invited Speaker, "Genes, Human Eye Disease, and Therapy," Bethesda, MD, 11/6/2006

American Academy of Ophthalmology (AAO) Annual Meeting, Symposium on Developing International Research Collaborations, Invited Speaker, "The View from the NEI," Las Vegas, NV, 11/13/2006

2007

Ocular Epidemiology Symposium, "Future Directions in Ocular Epidemiology," Sarasota, FL, 1/31/2007

Association of University Professors of Ophthalmology (AUPO) Annual Meeting, Speaker, "Update on NEI/NIH Activities," Indian Wells, CA, 2/3/2007

IOM Neuroscience Biomarkers & Biosignatures Workshop: Converging Technologies, Emerging Partnerships. Speaker, "Current and Near-term Impact of Biomarkers for Retinal Neurodegenerations," Washington DC, 2/26/2007

University of Michigan Genetics Grand Rounds, Speaker, "Dissecting a Mendelian Trait: X-linked Retinoschisis," Ann Arbor, MI, 3/8/2007

Unite for Sight, Stanford University, Invited Speaker, "A Global Perspective for Vision Research," Palo Alto, CA, 4/14/2007

The Association for Research in Vision and Ophthalmology 2007, Invited Speaker, "Ophthalmic Genetics: A Look to the Future of Medicine," Ft. Lauderdale, FL, 5/7/2007

The Association for Research in Vision and Ophthalmology 2007, Nanotechnology Group Organizing Conference, Speaker, "The NIH Nanomedicine Roadmap and the Vision Community," Ft. Lauderdale, FL, 5/8/2007

Maryland Society of Eye Physicians and Surgeons, Keynote Speaker, "Ophthalmic Genetics: A Look to the Future of Medicine," Baltimore, MD, 5/11/2007

Medical University of South Carolina, Storm Eye Institute, 8th Annual Kerrison Endowed Lecture, Featured Speaker, "Genetic Ophthalmology: A Look to the Future of Medicine," Charleston, SC, 6/14/2007

NIH Research Festival, 20th Annual, Symposium on Forestalling Blindness: Two Decades of Progress, Symposium Speaker, "Stemming Vision Loss with CNTF for Retinal Degenerative Disease," Bethesda, MD, 9/26/2007

National Institutes of Health, Medicine for the Public, 2007 Lecture Series, Invited Speaker, "From Childhood Blindness to Age-related Macular Degeneration: Genes, Eye Disease, and Prospects for Therapy," Bethesda, MD, 10/16/2007

Schepens International Society Meeting, Invited Speaker, "Therapeutic Strategies for Photoreceptor Neurodegenerative Retinal Dystrophies: CNTF Phase-1 Study for Retinitis Pigmentosa," Washington DC, 10/26/2007

Schepens International Society Meeting, Retina Symposium, Symposium Speaker, "Genetic Ophthalmology: A look to the Future of Medicine," Washington DC, 10/27/2007

Foundation for Fighting Blindness (FFB)/Alliance for Eye and Vision Research (AEVR), Congressional Briefing, "Rare Eye Diseases: On the Brink of Breakthroughs," Washington DC, 10/31/2007

American Acedemy of Ophthalmology (AAO), 2007 Annual Meeting, Invited Speaker, "The National Institutes of Health: Nanotechnology Roadmap," New Orleans, LA, 11/12/07

Prevention of Blindness Society of Metropolitan Washington, Saturday Sibley Hospital Lecture, Invited Speaker, "Ophthalmic Genetics: A Look to the Future of Medicine," Washington DC, 11/17/2007

2008

Association of University Professors of Ophthalmology (AUPO), Speaker, "Update on NEI/NIH Activities," Sarasota, FL, 2/2/2008

2008 Annual Meeting Association of Medical School Neuroscience Department Chairpersons, Invited Speaker, "Neuroscience at NIH and NEI," Christiansted, St. Croix, USVI, 3/7/2008

2008 Walter Reed Army Medical Center Ophthalmology Service Biennial Meeting: Current Treatments of Ocular Inflammation, Uveitis, Refractive Surgery, and Genetics, John Harry King Lecture, "Update on Ocular Genetics," Bethesda, MD, 3/24/2008

Case Western Reserve University, Visual Sciences Research Center Symposium, Invited speaker, "National Institutes of Health & National Eye Institute: Activities and Budget Perspectives," Cleveland, OH, 6/6/2008

Case Western Reserve University, Visual Sciences Research Center Symposium, Invited speaker, "Ophthalmic Genetics: Looking Toward Treatments," Cleveland, OH, 6/6/2008

Illinois Eye & Ear Infirmary 150th Anniversary, Department of Ophthalmology, University of Illinois, The Peter C. Kronfeld, M.D. Memorial Lecture, "Genetic Ophthalmology: Looking Towards Treatments," 6/20/2008

World Ophthalmology Congress. Invited Speaker. "Cell Based Drug Delivery to the Posterior Segment & the CNTF Implant," Hong Kong, China, 6/28/2008
World Ophthalmology Congress, Invited Speaker, "Beyond a National Perspective: Global Research to Decrease Visual Impairment and Blindness," Hong Kong, China, 6/29/2008
World Ophthalmology Congress, Invited Speaker, "Clinical Trial of Encapsulated CNTF for Retinitis Pigmentosa," Hong Kong, China, 6/30/2008
Extraordinary Ophthalmology Conference, University of Wisconsin, Invited speaker, "Genetic Ophthalmology: Heading for Treatments," Madison, WI, 7/11/2008
Extraordinary Ophthalmology Conference. University of Wisconsin, Invited speaker, "Nanotechnology, Nanomedicine, and Vision," Madison, WI, 7/11/2008
International Society for Clinical Electrophysiology of Vision (ISCEV) Annual Symposium, Invited Speaker, "Inner Retinal Sequelae of Outer retinal Disease: Origins of ERG Components in the RCS Rat," Morgantown, WV, 7/14/2008
International Society for Genetic Eye Diseases and Retinoblastoma (ISGEDR) Annual Meeting, Invited Speaker, "Gene Therapy for X-linked Retinoschisis," Strasbourg, France, 8/29/2008
XVIII International Congress for Eye Research Annual Meeting (ICER) Annual Meeting, Invited Speaker, "Background and Outcome of Phase-1 Human CNTF Clinical Trial for RP & Consideration of Phase-2 Design," Beijing, China, 9/28/2008
XVIII International Congress for Eye Research Annual Meeting (ICER) Annual Meeting, Co-chair and Speaker, "Ophthalmic Genetics: Looking Towards Treatments," Beijing, China, 9/28/2008
Annual Meeting of the American Academy Ophthalmology, Retina Subspecialty Day, Section VII: Inherited Retinal Diseases, "Phase I Clinical Safety Study for Human Retinitis Pigmentosa Using Ciliary Neurotrophic Factor (CNTF) Delivered by Sequestered Live Cell Intraocular Implants & Consideration of Phase II Design," Atlanta, GA, 11/2008
Vision Symposium—National d'Ophtalmologie des Quinze-Vingts, "Genetic Ophthalmology and Neuroscience: Heading for Treatments," Paris, France, 12/2008

2009

Asia/ARVO International Meeting on Research in Vision and Ophthalmology, Invited speaker, "Genetics and the Future of Ophthalmology," Hyderabad, India, 1/15/2009
Genetics of Eye Disease, Invited speaker, "Future Directions on the Genetics of Eye Disease: an Integrated Vision Across Genomics and Therapeutics," Catalina Island, CA, 2/4/2009
Gene Therapy and Eye Disease, "Therapy for Rare Retinal Degenerative Diseases," Invited Speaker, Rancho Santa Fe, CA, 6/15/2009
Vanderbilt Eye Institute, "Translational Research: Ophthalmic Genetics," Invited Speaker, Nashville, TN, 9/18/2009
Retina Congress 2009, "Integrated Vision Across Genomics, Bioengineering, and Therapeutics," Invited Speaker, New York, NY, 10/2/2009
AMA Ophthalmology Section Council Symposium, Joint Meeting of the American Academy of Ophthalmology and Pan-American Association of Ophthalmology, "Health Care Reform and Ophthalmologic Research," Invited Speaker, San Francisco, CA, 10/26/2009
University of Pittsburgh School of Medicine, Eye & Ear Institute, "Translational Research: Ophthalmic Genetics," Invited Speaker, Pittsburgh, PA, 11/6/2009

BIBLIOGRAPHY: LEGAL DOCUMENTS

1974 (NOTE: I wrote these as a summer intern with the Jerome Frank Legal Service Organization, Yale Law School, and they were submitted to the court under the direction of the supervising attorney.)

Petition for a Writ of Habeas Corpus: *Fiorenza v. Norton*, United States District Court of Connecticut, Civil No.B74–297.

Brief for Defendent-appellee: *Giordano v. Norton*, United States Court of Appeals Second Circuit, Docket No. 74–1394.

BIBLIOGRAPHY: JOURNAL ARTICLES

1978

Maggiano JM, Fishman GA, Evans LS, **Sieving PA**, Goldbaum M. Calibration error in dark adaptometer. *Arch Ophthalmol* 96:1082–3.

Sieving PA, Fishman GA. Refractive errors of retinitis pigmentosa patients. *Br J Ophthalmol* 62:163–7.

Sieving PA, Fishman GA, Maggiano JM. Corneal wick electrode for recording bright flash electroretinograms and early receptor potentials. *Arch Ophthalmol* 96:899–900.

1982

Sieving PA, Fishman GA. Rod contribution to the human ERP estimated from monochromats' data. *Doc Ophthalmol Proc Ser* 31:95–102.

1983

Sieving PA, Fishman GA, Alexander KR, Goldberg MF. Early receptor potential (ERP) measurements in human ocular siderosis. *Arch Ophthalmol* 101:1716–20.

1984

Jampol LM, **Sieving PA**, Pugh D, Fishman GA, Gilbert H. Multiple evanescent white dot syndrome—Part I: clinical findings. *Arch Ophthalmol* 102:671–4.

Sieving PA, Fishman GA, Salzano, Rabb MF. Acute macular neuroretinopathy: early receptor potential change suggests photoreceptor pathology. *Br J Ophthalmol* 68:229–34.

Sieving PA, Fishman GA, Jampol LM, Pugh D. Multiple evanescent white dot syndrome—Part II: electrophysiology of the photoreceptors during retinal pigment epithelial disease. *Arch Ophthalmol* 102:675–9.

1985

Sieving PA, Steinberg RH. Contribution from proximal retina to intraretinal pattern ERG: the m-wave. *Invest Ophthalmol Vis Sci* 26:1642–7.

1986

Sieving PA, Frishman LJ, Steinberg RH. M-wave of proximal retina in cat. *J Neurophysiol* 56(4):1039–48.

Sieving PA, Frishman LJ, Steinberg RH. Scotopic threshold response of proximal retina in cat. *J Neurophysiol* 56(4):1049–61.

Sieving PA, Niffenegger J, Berson EL. Electroretinographic findings in selected pedigrees with choroideremia. *Am J Ophthalmol* 101:361–7.

1987

Sieving PA, Steinberg RH. Proximal retinal contribution to the intraretinal 8-Hz pattern ERG of cat. *J Neurophysiol* 57(1):104–20.

1988

Frishman LJ, **Sieving PA**, Steinberg RH. Contributions to the electroretinogram of currents originating in proximal retina. *Vis Neurosci* 1:307–15.

Han D, **Sieving PA**, Johnson MW, Martonyi CL. Foveal retinoschisis associated with senile retinoschisis in a woman. *Am J Ophthalmol* 106:107–9.

Sieving PA, Nino C. Scotopic Threshold Response (STR) of the human electroretinogram. *Invest Ophthalmol Vis Sci* 29:1608–14.

Wakabayashi K, Gieser J, **Sieving PA**. Aspartate separation of the scotopic threshold response (STR) from the photoreceptor a-wave of the cat and monkey ERG. *Invest Ophthalmol Vis Sci* 29:1615–22.

1989

Ticho B, **Sieving P**. Leber's congenital amaurosis with marbleized fundus and juvenile nephronophthisis. *Am J Ophthalmol* 107:426–8.

1990

Rowe SE, Trobe JD, **Sieving PA**. Idiopathic photoreceptor dysfunction causes unexplained visual acuity loss in later adulthood. *Ophthalmology* 97:1632–7.

Sieving PA, Bingham EL, Roth MS, Young MR, Boehnke M, Kuo C-Y, Ginsburg D. Linkage relationship of X-linked juvenile retinoschisis with Xp22.1–p22.3 probes. *Am J Hum Genet* 47:616–21.

1991

Arrindell EL, Trobe JD, **Sieving PA**, Barnett JL. Pupillary and electroretinographic abnormalities in a family with neuronal intranuclear hyaline inclusion disease. *Arch Ophthalmol* 109:373–8.

Murayama K, Kuo C-Y, **Sieving PA**. Abnormal threshold ERG response in X-linked juvenile retinoschisis: evidence for a proximal retinal origin for the human STR. *Clin Vis Sci* 6:317–22.

Naarendorp F, **Sieving PA**. The scotopic threshold response of the cat ERG is suppressed selectively by GABA and glycine. *Vis Res* 31:1–15.

Richards JE, Kuo C-Y, Boehnke M, **Sieving PA**. Rhodopsin Thr58Arg mutation in a family with autosomal dominant retinitis pigmentosa. *Ophthalmology* 98:1797–805.

Sieving PA. Retinal ganglion cell loss does not abolish the scotopic threshold response (STR) of the cat and human ERG. *Clin Vis Sci* 6:149–58.

Sieving PA, Wakabayashi K. Comparison of rod threshold ERG from monkey, cat and human. *Clin Vis Sci* 6:171–9.

Sneed SR, **Sieving PA**. Fenestrated sheen macular dystrophy. *Am J Ophthalmol* 112:1–7.

1992

Geller AM, **Sieving PA**, Green DG. Effect on grating identification of sampling with degenerate arrays. *J Opt Soc Am* 9:472–7.

Murayama K, **Sieving PA**. Different rates of growth of monkey and human photopic a-, b- and d-waves suggests two sites of ERG light adaptation. *Clin Vis Sci* 7:385–92.

1993

Geller AM, **Sieving PA**. Assessment of foveal cone photoreceptors in Stargardt's macular dystrophy using a small dot detection task. *Vis Res* 33:1509–24.

Macke JP, Davenport CM, Jacobson SG, Hennessey JC, Gonzalez-Fernandez F, Heckenlively J, Finkelstein D, Palmer R, Maumenee IH, Pearce W, **Sieving P**, Gouras P, Nathans J. Identification of novel rhodopsin mutations responsible for retinitis pigmentosa: implications for the structure and function of rhodopsin. *Am J Hum Genet* 53:80–9.

Naarendorp F; Hitchcock PF; **Sieving PA**. Dopaminergic modulation of rod pathway signals does not affect the scotopic ERG of cat at dark-adapted threshold. *J Neurophysiol* 70:1681–91.

1994

Bush RA, **Sieving PA**. A proximal retinal component in the primate photopic ERG a-wave. *Invest Ophthalmol Vis Sci* 35:635–45.

Sieving PA. AOS Thesis: Photopic ON- and OFF-pathway abnormalities in retinal dystrophies. *Trans Am Ophthalmol Soc* LXXXXI:701–73.

Sieving PA. "Unilateral cone dystrophy": ERG changes implicate abnormal signaling by hyperpolarizing bipolar and/or horizontal cells. *Tr Am Ophth Soc* XCII:459–74.

Sieving PA, Murayama K, Naarendorp F. Push-pull model of the primate photopic electroretinogram: a role for hyperpolarizing neurons in shaping the b-wave. *Vis Neurosci* 11:519–32.

Yan D, Wong D, Zheng K, Thiselton D, Fujita R, **Sieving PA**, Bhattacharya SS, Yang-Feng TL, Richards JE, Swaroop A. Dinucleotide repeat polymorphism at the DXS977. *Hum Mol Genet* 3:1030.

1995

Bush RA, Hawks KW, **Sieving PA**. Preservation of inner retinal responses in the aged Royal College of Surgeons rat: evidence against glutamate excitotoxicity in photoreceptor degeneration. *Invest Ophthalmol Vis Sci* 36:2054–62.

Frishman LA, **Sieving PA**. Evidence for two sites of adaptation affecting the dark-adapted ERG of cats and primates. *Vis Res* 35:435–42.

Fujita R, Blumberg M, Anderson D, Forsythe P, McHenry C, Yan D, Yang-Feng TL, **Sieving PA**, Swaroop A. A polymorphic trinucleotide repeat at *DXS8170* in the critical region of X-linked retinitis pigmentosa locus *RP3* at Xp21.1. *Mol Vis* 1:3.

Pawar H, Bingham EL, Lunetta KL, Segal M, Richards J, Boehnke M, **Sieving PA**. Refined genetic mapping of juvenile X-linked retinoschisis. *Hum Hered* 45:206–10.

Richards JE, Scott KM, **Sieving PA**. Disruption of conserved rhodopsin disulfide bond by Cys187Tyr mutation causes early and severe autosomal dominant retinitis pigmentosa. *Ophthalmology* 102:669–77.

Sieving, PA, Richards JE, Naarendorp F, Bingham EL, Scott K, Alpern M. Dark-light: model for night-blindness from the human rhodopsin Gly90→Asp mutation. *Proc Natl Acad Sci (USA)* 92:880–4.

1996

Bush RA, **Sieving PA**. Inner retinal contributions to the primate photopic fast flicker electroretinogram. *J Opt Soc Am* 13:557–65.

Fujita R, Bingham E, Forsythe P, McHenry C, Aita V, Bradford AN, Dry K, Segal M, Devoto M, Bruns G, Wright AF, Ott J, **Sieving PA**, Swaroop A. A recombination outside the *BB* deletion refines the location of the X-linked retinitis pigmentosa locus *RP3*. *Am J Hum Genet* 59:152–8.

Hou Y-C, Richards JE, Bingham E, Pawar H, Scott K, Segal M, Lunetta KL, Boehnke M, **Sieving PA**. Linkage study of Best's vitelliform macular dystrophy (VMD2) in a large North American family. *Hum Hered* 46:211–20.

Naarendorp F, Rice KS, **Sieving PA**. Summation of rod and S-cone signals at threshold in human observers. *Vision Res* 36:2681–8.

Pawar H, Bingham EL, Hiriyanna K, Segal M, Richards JE, **Sieving PA**. X-linked juvenile retinoschisis: localization between (DXS1195, DXS418) and AFM291wf5 on a single YAC. *Hum Hered* 46:329–35.

Sieving PA, Boskovich S, Bingham E, Pawar, H. Sorsby's fundus dystrophy in a family with a SER-181-CYS mutation in the TIMP-3 gene: poor outcome after laser photocoagulation. *Tr Am Ophth Soc*, XCIV: 275–97.

1997

Buraczynska M, Wu W, Fujita R, Buraczynska K, Phelps E, Andreasson S, Bennett J, Birch DG, Fishman GA, Hoffman DR, Inana G, Jacobson SG, Musarella MA, **Sieving PA**, Swaroop A. Spectrum of mutations in the *RPGR* gene that are identified in 20% of families with X-linked retinitis pigmentosa. *Am J Hum Gen* 61:1287–92.

Farjo Q, Jackson A, Pieke-Dahl S, Scott K, Kimberling WJ, **Sieving PA**, Richards JE, Swaroop A. Human bZIP transcription factor gene *NRL*: structure, genomic sequence, and fine linkage mapping at 14q11.2 and negative mutation analysis in patients with retinal degeneration. *Genomics* 45:395–401.

Fujita R, Buraczynska M, Gieser L, Wu W, Forsythe P, Abrahamson M, Jacobson SG, **Sieving PA**, Andreasson S, Swaroop A. Analysis of the *RPGR* gene in 11 pedigrees with the Retinitis Pigmentosa Type 3 genotype: paucity of mutations in the coding region but splice defects in two families. *Am J Hum Gen* 61:571–80.

Humphries MM, Rancourt D, Farrar GJ, Kenna P, Hazel M, Bush RA, **Sieving PA**, Sheils DM, McNally N, Creighton P, Erven A, Boros A, Gulya K, Capecchi MR, Humphries P. Retinopathy induced in mice by targeted disruption of the rhodopsin gene. *Nat Genet* 15:216–9.

Kim S, Bush RA, **Sieving PA**. Increased phase lag of the fundamental harmonic component of the 30-Hz flicker ERG in Schubert-Bornschein complete-type CSNB. *Vis Res* 37:2471–5.

Swain PK, Chen S, Wang Q-L, Affatigato LM, Coats CL, Brady KD, Fishman GA, Jacobson SG, Swaroop A, Stone E, **Sieving PA**, Zack DJ. Mutations in the cone-rod homeobox gene are associated with the cone-rod dystrophy photoreceptor degeneration. *Neuron* 19:1329–36.

1998

Innis JW, **Sieving PA**, McMillan P, Weatherly RA. Apparently new syndrome of sensorineural hearing loss, retinal pigment epithelium lesions, and discolored teeth. *Am J Med Gen* 75:13–7.

Retinoschisis Consortium, The *(Group 3:* Hiriyanna KT, Bingham EL, McHenry C, Pawar H, Coats C, Darga T, Richards JE, **Sieving PA**: W. K. Kellogg Eye Center, University of Michigan, Ann Arbor, MI, USA). Functional implications of the spectrum of mutations found in 234 cases with X-linked juvenile retinoschisis (XLRS). *Hum Mol Genet* 7:1185–92.

Sieving PA, Arnold EB, Jamison J, Liepa A, Coats C. Submicrovolt flicker electroretinogram: cycle-by-cycle recording of multiple harmonics with statistical estimation of measurement uncertainty. *Invest Ophthalmol Vis Sci* 39:1462–9.

Sugawara T, **Sieving PA**, Iuvone M, Bush RA. The melatonin antagonist luzindole protects retinal photoreceptors from light damage in the rat. *Invest Ophthalmol Vis Sci* 39:2458–65.

1999

Ayyagari R, Kakuk LE, Coats CL, Bingham EL, Toda Y, Felius J, **Sieving PA**. Bilateral macular atrophy in blue cone monochromacy (BCM) with loss of the locus control region (LCR) and part of the red pigment gene. *Mol Vis* 5:13.

Caldwell GM, Kakuk LE, Griesinger IB, Simpson SA, Nowak NJ, Small KW, Maumenee IH, Rosenfeld PJ, **Sieving PA**, Shows TB, Ayyagari R. Bestrophin gene mutations in patients with Best vitelliform macular dystrophy. *Genomics* 58:98–101.

Hiriyanna KT, Bingham EL, Yashar BM, Ayyagari R, Fishman G, Small KW, Weinberg DV, Weleber RG, Lewis RA, Andreasson S, Richards JE, **Sieving PA**. Novel mutations in XLRS1 causing retinoschisis, including first evidence of putative leader sequence change. *Hum Mut* 14:423–7.

McNally N, Kenna P, Humphries MM, Hobson AH, Khan NW, Bush RA, **Sieving PA**, Humphries P, Farrar GJ. Structural and functional rescue of murine rod photoreceptors by human rhodopsin transgene. *Hum Mol Genet* 8:1309–12.

Mears AJ, Gieser L, Yan D, Chen C, Fahrner S, Hiriyanna S, Fujita R, Jacobson SG, **Sieving PA**, Swaroop A. Protein-truncation mutations in the *RP2* gene in a North American cohort of families with X-linked retinitis pigmentosa. *Am J Hum Gen* 64:897–900.

Mendoza-Londono R, Hiriyanna KT, Bingham EL, Rodriguez F, Shastry BS, Rodriguez A, **Sieving PA**, Tamayo ML. A Colombian family with X-linked juvenile retinoschisis with three affected females: finding of a frameshift mutation. *Ophthalmic Genet* 20:37–43.

Sieving PA, Bingham EL, Kemp J, Richards J, Hiriyanna K. Juvenile X-linked retinoschisis from *XLRS1* Arg213Trp mutation with preservation of the electroretinogram scotopic b-wave. *Am J Ophthalmol* 128:179–84.

Sieving PA, Yashar BM, Ayyagari R. Juvenile retinoschisis: a model for molecular diagnostic testing of X-linked ophthalmic disease. *Trans Am Ophthalmol Soc* 97:451–64.

Swaroop A, Wang Q-L, Wu W, Cook J, Coats C, Xu S, Chen S, Zacks DJ, **Sieving PA**. Leber congenital amaurosis caused by a homozygous mutation (R90W) in the homeodomain of the retinal transcription factor CRX: direct evidence for the involvement of CRX in the development of photoreceptor function. *Hum Mol Genet* 8:299–305.

Toda K, Bush RA, Humphries P, **Sieving PA**. The electroretinogram of the rhodopsin knockout mouse. *Vis Neurosci* 16:391–8.

Walpole SM, Hiriyanna KT, Nicolaou A, Bingham EL, Durham J, Vaudin M, Ross MT, Yates JR, **Sieving PA**, Trump D. Identification and characterization of the human homologue (*RAI2*) of a mouse retinoic acid-induced gene in Xp22. *Genomics* 55:275–83.

2000

Ayyagari R, Griesinger IB, Bingham E, Lark KK, Moroi SE, **Sieving PA**. Autosomal dominant hemorrhagic macular dystrophy not associated with the *TIMP3* Gene. *Arch Ophthalmol* 118:85–92.

Ayyagari R, Kakuk LE, Bingham EL, Szczesny JJ, Kemp J, Toda Y, Felius J, **Sieving PA**. Spectrum of color gene deletions and phenotype in patients with blue cone monochromacy. *Hum Gen* 107:75–82.

Bush RA, Kononen L, Machida S, **Sieving PA**. The effect of calcium channel blocker diltiazem on photoreceptor degeneration in the rhodopsin *pro23His* rat. *Invest Ophthalmol Vis Sci* 41:2697–701.

Eksandh LC, Ponjavic V, Ayyagari R, Bingham EL, Hiriyanna KT, Andreasson S, Ehinger B, **Sieving PA**. Phenotypic expression of juvenile X-linked retinoschisis in Swedish families with different mutations in the *XLRS1* gene. *Arch Ophthalmol* 118:1098–104.

Griesinger IB, **Sieving PA**, Ayyagari R. Autosomal dominant macular atrophy at 6q14 excludes CORD7 and MCDR1/PBCRA loci. *Invest Ophthalmol Vis Sci* 41:248–55.

Hrach CJ, Johnson MW, Hassan AS, Lei B, **Sieving PA**, Elner VM. Retinal toxicity of commercial intravitreal tissue plasminogen activator solution in cat eyes. *Arch Ophthalmol* 118:659–63.

Kohl S, Baumann B, Broghammer M, Jägle H, **Sieving PA**, Kellner U, Spegal R, Anastasi M, Zrenner E, Sharpe LT, Wissinger B. Mutation in the *CNGB3* gene encoding the β-subunit of the cone photoreceptor cGMP-gated channel are responsible for achromatopsia (*ACHM3*) linked to chromosome 8q21. *Hum Mol Genet* 9:2107–16.

Lei B, Bush RA, Milam AH, **Sieving PA**. Human melanoma-associated retinopathy (MAR) antibodies alter the retinal ON-response of the monkey ERG *in vivo*. *Invest Ophthalmol Vis Sci* 41:262–6.

Machida S, Kondo M, Jamison J, Khan NW, Kononen LT, Sugawara T, Bush RA, **Sieving PA**. P23H rhodopsin transgenic rat: correlation of retinal function with histopathology. *Invest Ophthalmol Vis Sci* 41:3200–9.

Mears AJ, Hiriyanna S, Vervoort R, Yashar B, Gieser L, Fahrner S, Daiger SP, Heckenlively JR, **Sieving PA**, Wright AF, Swaroop A. Remapping of the *RP15* locus for X-linked cone-rod degeneration to Xp11.4–p21.1, and identification of a de novo insertion in the *RPGR* exon ORF15. *Am J Hum Gen* 67:1000–3.

Sugawara T, **Sieving PA**, Bush RA. Quantitative relationship of the scotopic and photopic ERG to photoreceptor cell loss in light damaged rats. *Exp Eye Res* 70:693–705.

Thompson DA, Gyürüs P, Fleischer LL, Bingham EL, McHenry CL, Apfelstedt-Sylla E, Zrenner E, Lorenz B, Richards JE, Jacobson SG, **Sieving PA**, Gal A. Genetics and phenotypes of *RPE65* mutations in inherited retinal degeneration. *Invest Ophthalmol Vis Sci* 41:4293–9.

2001

Ayyagari R, Zhang K, Hutchinson A, Yu Z, Swaroop A, Kakuk LE, Seddon JM, Bernstein PS, Lewis RA, Tammur J, Yang Z, Li Y, Zhang H, Yashar BM, Liu J, Petrukhin K, **Sieving PA**, Allikmets R. Evaluation of the ELOVL4 gene in patients with age-related macular degeneration. *Ophthalmic Genet* 22:233–9.

Humphries MM, Kiang S, McNally N, Donovan MA, **Sieving PA**, Bush RA, Machida S, Cotter T, Hobson A, Farrar J, Humphries P, Kenna P. Comparative structural and functional analysis of photoreceptor neurons of *Rho*-/- mice reveal increased survival on C57BL/6J in comparison to 129Sv genetic background. *Vis Neurosci* 18: 437–43.

Jamison JA, Bush RA, Lei B, **Sieving PA**. Characterization of the rod photoresponse isolated from the dark-adapted primate ERG. *Vis Neurosci* 18:445–55.

Khan NW, Jamison JA, Kemp JA, **Sieving PA**. Analysis of photoreceptor function and inner retinal activity in juvenile X-linked retinoschisis. *Vision Res* 41:3931–42.

Kondo M, **Sieving PA**. Primate photopic sine-wave flicker ERG: vector modeling analysis of component origins using glutamate analogs. *Invest Ophthalmol Vis Sci* 42:305–12.

Machida S, Chaudhry P, Shinohara T, Singh DP, Reddy VN, Chylack LT Jr, **Sieving PA**, Bush RA. Lens epithelium-derived Growth Factor promotes photoreceptor survival in light-damaged and RCS rats. *Invest Ophthalmol Vis Sci* 42:1087–95.

Mears AJ, Kondo M, Swain PK, Takada Y, Bush RA, Saunders TL, **Sieving PA**, Swaroop A. Nrl is required for rod photoreceptor development. *Nat Genet* 29:447–52.

Sieving PA. Fifteen years of work: the COMS outcomes for medium-sized choroidal melanoma. *Arch Ophthalmol* 119: 1067–8.

Sieving PA, Chaudhry P, Kondo M, Provenzano M, Wu D, Carlson TJ, Bush RA, Thompson DA. Inhibition of the visual cycle *in vivo* by 13-*cis* retinoic acid protects from light damage and provides a mechanism for night blindness in isotretinoin therapy. *Proc Natl Acad Sci (USA)* 98:1835–40.

Sieving PA, Fowler ML, Bush RA, Machida S, Calvert PD, Green DG, Makino CL, McHenry CL. Constitutive "light" adaptation in rods from G90D rhodopsin: a mechanism for human congenital night blindness without rod cell loss. *J Neurosci* 21:5449–60.

Thompson DA, Li Y, McHenry CL, Carlson TJ, Ding X, **Sieving PA**, Apfelstedt-Sylla E, Gal A. Mutations in the gene encoding lecithin retinol acyltransferase are associated with early-onset severe retinal dystrophy. *Nat Genet* 28: 123–4.

Weinberg DV, **Sieving PA**, Bingham EL, Jampol LM, Mets MB. Bietti crystalline retinopathy and juvenile retinoschisis in a family with a novel RS1 mutation (Letter to Editor*). Arch Ophthalmol* 119:1719 –21.

Zhang K, Kniazeva M, Han M, Li W, Yu Z, Yang Z, Li Y, Metzker ML, Allikmets R, Zack DJ, Kakuk LE, Lagali PS, Wong PW, MacDonald IM, **Sieving PA**, Figueroa DJ, Austin CP, Gould RJ, Ayyagari R, Petrukhin K. A 5-bp deletion in *ELOVL4* is associated with two related forms of autosomal dominant macular dystrophy. *Nat Genet* 27:89–93.

2002

Ayyagari R, Demirci FY, Liu J, Bingham EL, Stringham H, Kakuk LE, Boehnke M, Gorin MB, Richards JE, **Sieving PA**. X-linked recessive atrophic macular degeneration from *RPGR* mutation. *Genomics* 80: 166–71.

Breuer DK, Yashar BM, Filippova E, Hiriyanna S, Lyons RH, Mears AJ, Asaye B, Acar C, Vervoort R, Wright AF, Musarella MA, Wheeler P, MacDonald I, Iannaccone A, Birch D, Hoffman DR, Fishman GA, Heckenlively JR, Jacobson SG, **Sieving PA**, Swaroop A. A comprehensive mutation analysis of *RP2* and *RPGR* in a North American cohort of families with X-linked retinitis pigmentosa. *Am J Hum Genet* 70:1545–54.

Felius J, Thompson DA, Khan NW, Bingham EL, Jamison JA, Kemp JA, **Sieving PA**. Clinical course and visual function in a family with mutations in the *RPE65* gene. *Arch Ophthalmol* 120:55–61.

Kondo M, **Sieving PA**. Post-photoreceptoral activity dominates primate photopic 32-Hz ERG for sine-, square-, and pulsed stimuli. *Invest Ophthalmol Vis Sci* 43:2500–7.

Traverso V, Bush RA, **Sieving PA**, Deretic D. Retinal cAMP levels during the progression of retinal degeneration in rhodopsin P23H and S334ter transgenic rats. *Invest Ophthalmol Vis Sci* 43:1655–61.

2003

Acar C, Mears AJ, Yashar BM, Maheshwary AS, Andreasson S, Baldi A, **Sieving PA**, Iannaccone A, Musarella MA, Jacobson SG, Swaroop A. Mutation screening of patients with Leber congenital amaurosis or the enhanced S-cone syndrome reveals a lack of sequence variations in the *NRL* gene. *Mol Vis* 9:14–7.

Ahmed ZM, Riazuddin S, Ahmad J, Bernstein SL, Guo Y, Sabar MF, **Sieving P**, Griffith AJ, Friedman TB, Belyantseva IA, Wilcox ER. *PCDH15* is expressed in the neurosensory epithelium of the eye and ear and mutant alleles are responsible for both USH1F and DFNB23. *Hum Mol Genet* 12:3215–23.

Lee TK, McTaggart KE, **Sieving PA**, Heckenlively JR, Levin AV, Greenberg J, Weleber RG, Tong PY, Anhalt EF, Powell BR, MacDonald IM. Clinical diagnoses that overlap with choroideremia. *Can J Ophthalmol* 38:364–72; quiz 372.

Moroi SE, Lark KK, **Sieving PA**, Nouri-Mahdavi K, Schlotzer-Schrehardt U, Katz GJ, Ritch R. Long anterior zonules and pigment dispersion. *Am J Ophthalmol* 136:1176–8.

Radu RA, Mata NL, Nusinowitz S, Liu X, **Sieving PA**, Travis GH. Treatment with isotretinoin inhibits lipofuscin accumulation in a mouse model of recessive Stargardt's macular degeneration. *Proc Natl Acad Sci (USA)* 100: 4742–7.

Rivolta C, Ayyagari R, **Sieving PA**, Berson EL, Dryja TP. Evaluation of the *ELOVL4* gene in patients with autosomal recessive retinitis pigmentosa and Leber congenital amaurosis. *Mol Vis* 9:49–51.

Schimmenti LA, Manligas GS, **Sieving PA**. Optic nerve dysplasia and renal insufficiency in a family with a novel *PAX2* mutation, Arg115X: further ophthalmologic delineation of the renal-coloboma syndrome. *Ophthalmic Genet* 24:191–202.

2004

Abecasis GR, Yashar BM, Zhao Y, Ghiasvand NM, Zareparsi S, Branham KE, Reddick AC, Trager EH, Yoshida S, Bahling J, Filippova E, Elner S, Johnson MW, Vine AK, **Sieving PA**, Jacobson SG, Richards JE, Swaroop A. Age-related macular degeneration: a high resolution genome scan for susceptibility loci in a population enriched for late-stage disease. *Am J Hum Genet* 74:482–94.

Ambasudhan R, Wang XF, Jablonski MM, Thompson DA, Lagali PS, Wong PW, **Sieving PA**, Ayyagari, R. Atrophic macular degeneration mutations in *ELOVL4* result in the intracellular misrouting of the protein. *Genomics* 83:615–25.

Bush RA, Lei B, Tao W, Raz D, Chan C, Cox TA, Santos-Muffley M, **Sieving PA**. Encapsulated cell-based intraocular delivery of ciliary neurotrophic factor in normal rabbit: dose-dependent effects on ERG and retinal histology. *Invest Ophthalmol Vis Sci* 45(7):2420–30.

Mandal NA, Ambasudhan R, Wong PW, Gage PJ, **Sieving PA**, Ayyagari R. Characterization of mouse orthologue of *ELOVL4*: genomic organization and spatial and temporal expression. *Genomics* 83:626–35.

McHenry CL, Liu Y, Feng W, Nair AR, Feathers KL, Ding X, Gal A, Vollrath D, **Sieving PA**, Thompson DA. *MERTK* arginine-844-cysteine in a patient with severe rod-cone dystrophy: loss of mutant protein function in transfected cells. *Invest Ophthalmol Vis Sci* 45(5):1456–63.

Takada Y, Fariss RN, Tanikawa A, Zeng Y, Carper D, Bush R, **Sieving PA**. A retinal neuronal developmental wave of retinoschisin expression begins in ganglion cells during layer formation. *Invest Ophthalmol Vis Sci* 45(9):3302–12.

Zeng Y, Takada Y, Kjellstrom S, Hiriyanna K, Tanikawa A, Wawrousek E, Smaoui N, Caruso R, Bush RA, **Sieving PA**. *RS-1* gene delivery to an adult *Rs1h* knockout mouse model restores ERG b-wave with reversal of the electronegative waveform of X-linked retinoschisis. *Invest Ophthalmol Vis Sci* 45(9):3279–85.

2005

Ayyagari R, Mandal MN, Karoukis AJ, Chen L, McLaren NC, Lichter M, Wong DT, Hitchcock PF, Caruso RC, Moroi SE, Maumenee IH, **Sieving PA**. Late-onset macular degeneration and long anterior lens zonules result from a CTRP5 gene mutation. *Invest Ophthalmol Vis Sci* 46:3363–71.

Brzezinski JA 4th, Brown NL, Tanikawa A, Bush RA, **Sieving PA**, Vitaterna MH, Takahashi JS, Glaser T. Loss of circadian photoentrainment and abnormal retinal electrophysiology in math5 mutant mice. *Invest Ophthalmol Vis Sci* 46:2540–51.

Khan NW, Kondo M, Hiriyanna KT, Jamison JA, Bush RA, **Sieving PA**. Primate retinal signaling pathways: suppressing ON-pathway activity in monkey with glutamate analogs mimics human CSNB1-*NYX* genetic night blindness. *J Neurophysiol* 93:481–92.

Kohl S, Varsanyi B, Abadin GA, Baumann B, Hoyng C, Jägle H, Rosenberg T, Kellner U, Lorenz B, Salati R, Jurklies B, Farkas A, Andreasson S, Weleber R, Jacobson SG, Rudolph G, Castellan C, Dollfus H, Legius E, Anastasi M, Bitoun P, Lev D, **Sieving PA**, Munier FL, Zrenner E, Sharp LT, Cremers FPM, Wissinger B. *CNGB3* mutations account for 50% of all cases with autosomal recessive achromatopsia. *Eur J Hum Genet* 13:302–8.

Mandal MN, Heckenlively JR, Burch T, Chen L, Vasireddy V, Koenekoop RK, **Sieving PA**, Ayyagari R. Sequencing arrays for screening multiple genes associated with early-onset human retinal degenerations on a high-throughput platform. *Invest Ophthalmol Vis Sci* 46(9):3355–62.

Riazuddin SA, Zulfiqar F, Zhang Q, Sergeev YV, Qazi ZA, Husnain T, Caruso R, Riazuddin S, **Sieving PA**, Hejtmancik JF. Autosomal recessive retinitis pigmentosa is associated with mutations in RP1 in three consanguineous Pakistani families. *Invest Ophthalmol Vis Sci* 46(7):2264–70.

Thompson DA, Janecke AR, Lange J, Feathers KL, Hübner CA, McHenry CL, Stockton DW, Rammesmayer G, Lupski JR, Antinolo G, Ayuso C, Baiget M, Gouras P, Heckenlively JR, den Hollander A, Jacobson SG, Lewis RA, **Sieving PA**, Wissinger B, Yzer S, Zrenner E, Utermann G, and Gal A. Retinal degeneration associated with *RDH12* mutations results from decreased 11-*cis* retinal synthesis due to disruption of the visual cycle. *Hum Mol Genet* 14(24):3865–75.

Vasireddy V, Vijayasarathy C, Huang J, Wang XF, Jablonski MM, Petty HR, **Sieving PA**, Ayyagari, R. Stargardt-like macular dystrophy protein ELOVL4 exerts a dominant negative effect by recruiting wild-type protein into aggresomes. *Mol Vis* 11:665–76.

Zhang Q, Zulfiqar F, Riazuddin SA, Xiao X, Yasmeen A, Rogan PK, Caruso R, **Sieving PA**, Riazuddin S, Hejtmancik JF. A variant form of Oguchi disease mapped to 13q34 associated with partial deletion of *GRK1* gene. *Mol Vis* 11:977–85.

Zhang Q, Zulfiqar F, Xiao X, Riazuddin SA, Ayyagari R, Sabar F, Caruso R, **Sieving PA**, Riazuddin S, Hejtmancik JF. Severe autosomal recessive retinitis pigmentosa maps to chromosome 1p13.3–p21.2 between D1S2896 and D1S457 but outside ABCA4. *Hum Genet* 118:356–65.

2006

Aslanukov A, Bhowmick R, Guruju M, Oswald J, Raz D, Bush RA, **Sieving PA**, Lu X, Bock CB, Ferreira PA. RanBP2 modulates Cox11 and hexokinase I activities and haploinsufficiency of *RanBP2* causes deficits in glucose metabolism. *PLoS Genet* 2(10): 1653–65; e177.

Camasamudrum V, Gawinowicz MA, Zeng Y, Takada Y, Bush RA, **Sieving PA**. Identification and characterization of two mature isoforms of retinoschisin in murine retina. *Biochem Biophys Res Commun* 349(1):99–105.

Haywood-Watson RJ 2nd, Ahmed ZM, Kjellstrom S, Bush RA, Takada Y, Hampton LL, Battey JF, **Sieving PA**, Friedman TB. Ames Waltzer deaf mice have reduced electroretinogram amplitudes and complex alternative splicing of Pcdh15 transcripts. *Invest Ophthalmol Vis Sci* 47(7):3074–84.

Prenner JL, Capone A Jr, Ciaccia S, Takada Y, **Sieving PA**, Trese MT. Congenital X-linked retinoschisis classification system. *Retina* 26/7:S61–4.

Raz-Prag D, Ayyagari R, Fariss RN, Mandal MN, Vasireddy V, Majchrzak S, Webber AL, Bush RA, Salem N Jr, Petrukhin K, **Sieving PA**. Haploinsufficiency is not the key mechanism of pathogenesis in a heterozygous *Elovl4* knockout mouse model of STGD3 disease. *Invest Ophthal Vis Sci* 47(8):3603–11.

Riazuddin SA, Zulfiqar F, Zhang Q, Yao W, Li S, Jiao X, Shahzadi A, Amer M, Iqbal M, Hussnain T, **Sieving PA**, Riazuddin S, Hejtmancik JF. Mutations in the gene encoding the α-subunit of rod phosphodiesterase in consanguineous Pakistani families. *Mol Vis* 12:1283–91.

Sieving PA, Caruso RC, Tao W, Coleman HR, Thompson DJS, Fullmer KR, Bush RA. Ciliary neurotrophic factor (CNTF) for human retinal degeneration: phase-1 trial of CNTF delivered by encapsulated cell intraocular implants. *Proc Natl Acad Sci (USA)* 103(10):3896–3901.

Takada Y, Fariss RN, Müller M, Bush RA, Rushing EJ, **Sieving PA**. Retinoschisin expression and localization in rodent and human pineal and consequences of mouse *RS1* gene knockout. *Mol Vis* 12:1108–16.

Vasireddy V, Jablonski MM, Mandal MN, Raz-Prag D, Wang XF, Nizol L, Iannaccone A, Musch DC, Bush RA, Salem N Jr, **Sieving PA**, Ayyagari R. *Elovl4* 5-bp-deletion knock-in mice develop progressive photoreceptor degeneration. *Invest Ophthalmol Vis Sci* 47(10):4558–68.

Wen R, Song Y, Kjellstrom S, Tanikawa A, Liu Y, Li Y, Zhao Y, Bush RA, Laties AM, **Sieving PA**. Regulation of rod phototransduction machinery by ciliary neurotrophic factor. *J Neurosci* 26(52):13523–30.

2007

Downs K, Zacks DN, Caruso R, Karoukis AJ, Branham K, Yashar BM, Haimann M, Trzupek K, Meltzer M, Blain D, Richards JE, Weleber RG, Heckenlively JR, **Sieving PA**, Ayyagari R. Molecular testing for hereditary retinal disease as part of clinical care. *Arch Ophthalmol*, 125(2):252–8.

Khan NW, Wissinger B, Kohl S, **Sieving PA**. *CNGB3* achromatopsia with progressive loss of residual cone function and impaired rod-mediated function. *Invest Ophthalmol Vis Sci* 48(8):3864–71.

Kjellstrom S, Bush RA, Zeng Y, Takada Y, **Sieving PA**. Retinoschisin gene therapy and natural history in the *Rs1h*-KO mouse: long-term rescue from retinal degeneration. *Invest Ophthal Vis Sci* 48(8): 3837–45.

MacDonald IM, Brooks BP, **Sieving PA**. Eyeing a new network. *Science* 318(5853):1068.

MacDonald IM, Sauvé Y, **Sieving PA**. Preventing blindness in retinal disease: ciliary neurotrophic factor intraocular implants. *Can J Ophthalmol* 42(3):399–402.

Sieving PA, Collins FS. Genetic ophthalmology and the era of clinical care. *JAMA* 297(7): 733–6.

Vijayasarathy C, Takada Y, Zeng Y, Bush RA, **Sieving PA**. Retinoschisin is a peripheral membrane protein with affinity for anionic phospholipids and affected by divalent cations. *Invest Ophthalmol Vis Sci* 48(3): 991–1000.

Woodruff ML, Olshevskaya EV, Savchenko AB, Peshenko IV, Barrett R, Bush RA, **Sieving PA**, Fain GL, Dizhoor AM. Constitutive excitation by Gly90Asp rhodopsin rescues rods from degeneration caused by elevated production of cGMP in the dark. *J Neurosci* 27(33):8805–15.

Zhang Q, Zulfiqar F, Xiao X, Riazuddin SA, Ahmad Z, Caruso R, MacDonald I, **Sieving PA**, Riazuddin S, Hejtmancik JF. Severe retinitis pigmentosa mapped to 4p15 and associated with a novel mutation in the PROM1 gene. *Hum Genet.* 122(3–4):293–9.

2008

Ahmed ZM, Kjellstrom S, Haywood-Watson RJ, Bush RA, Hampton LL, Battey JF, Riazuddin S, Frolenkov G, **Sieving PA**, Friedman TB. Double homozygous waltzer and Ames waltzer mice provide no evidence of retinal degeneration. *Mol Vis* 14:2227–36.

Brooks BP, Macdonald IM, Tumminia SJ, Smaoui N, Blain D, Nezhuvingal AA, **Sieving PA**. Genomics in the era of molecular ophthalmology: reflections on the National Ophthalmic Disease Genotyping Network (eyeGENE). *Arch Ophthalmol* 126(3):424–5.

Dizhoor AM, Woodruff ML, Olshevskaya EV, Cilluffo MC, Cornwall MC, **Sieving PA**, Fain GL. Night blindness and the mechanism of constitutive signaling of mutant G90D rhodopsin. *J Neurosci* 28(45): 11662–72.

Machida S, Raz-Prag D, Fariss RN, **Sieving PA**, Bush RA. Photopic ERG negative response from amacrine cell signaling in RCS rat retinal degeneration. *Invest Ophthalmol Vis Sci* 49(1):442–52.

Takada Y, Vijayasarathy C, Zeng Y, Kjellstrom S, Bush RA, **Sieving PA**. Synaptic pathology in retinoschisis knockout (*Rs1*$^{-/y}$) mouse retina and modification by rAAV-*Rs1* gene delivery. *Invest Ophthalmol Vis Sci* 49(8):3677–86.

Vijayasarathy C, Takada Y, Zeng Y, Bush RA, **Sieving PA**. Organization and molecular interactions of retinoschisin in photoreceptors. *Adv Exp Med Biol* 613:291–7.

2009

Haruta M, Bush RA, Kjellstroma S, Vijayasarathya C, Zeng Y, Le Y-Z, **Sieving PA**. Depleting Rac1in mouse rod photoreceptors protects them from photo-oxidative stress without affecting their structure or function. *Proc Natl Acad Sci (USA)* 106(23):9397–9402.

Luna G, Kjellstrom S, Verardo M, Lewis GP, Byun J, **Sieving PA**, Fisher SK. The effects of transient retinal detachment on cavity size and glial and neural remodeling in a mouse model of X-linked retinoschisis. *Invest Ophthalmol Vis Sci* 50(8):3977-84.

Park TK, Wu Z, Kjellstrom S, Zeng Y, Bush RA, **Sieving PA**, Colosi P. Intravitreal delivery of AAV8 retinoschisin results in cell type-specific gene expression and retinal rescue in the *RS1*-Ko mouse. *Gene Therapy* 16:916–26.

Raz-Prag D, Zeng Y, **Sieving PA**, Bush RA. Photoreceptor protection by adeno-associated virus-mediated LEDGF expression in the RCS rat model of retinal degeneration: probing the mechanism. *Invest Ophthalmol Vis Sci* 50(8):3897–906.

Vijayasarathy C, Ziccardi L, Zeng Y, Smaoui N, Caruso RC, **Sieving PA**. Null retinoschisin-protein expression from an *RS1* c354del1-ins18 mutation causes progressive and severe XLRS in a cross-sectional family study. *Invest Ophthalmol Vis Sci.* 50(11):5375-83.

2010

Sieving PA. At the frontier of vision research—the National Eye Institute celebrates 40 years. *Am J Ophthamol* 149(2):179-81.

BIBLIOGRAPHY: BOOK CHAPTERS

1991

Steinberg RH, Frishman LJ, **Sieving PA**. Negative components of the electroretinogram from proximal retina and photoreceptor. In: *Progress in Retinal Research*, Pergamon Press, New York, Vol 10:121–160. 1991.

Sieving PA. In: *Handbook of Clinical Electrophysiology of Vision Testing* (eds.: J Heckenlively, G Arden). Mosby Year Book Publishers, St. Louis, 1991.

Chap. 26: Digital band-pass filtering for electrophysiologic recording systems, 205–10.

Chap. 46: Scotopic threshold response, 352–62.

Chap. 101: Electroretinographic detection of female carriers (Heterozygotes) of X-linked recessive retinitis pigmentosa, 741–3.

Chap. 102: Evaluation of the X-linked carrier state in choroideremia, 744–7.

1993

Sieving PA, Murayama K. Retinitis Pigmentosa. Chap. 65, 534–41. In: *Diagnostic Problems in Clinical Ophthalmology* (eds.: C Margo, LM Hamed, RN Mames). WB Saunders, Philadelphia, 1993.

Sieving PA, Wakabayashi K. Bull's-eye Maculopathies. Chap. 67, 547–55. In: *Diagnostic Problems in Clinical Ophthalmology* (eds.: C Margo, LM Hamed, RN Mames). WB Saunders, Philadelphia, 1993.

Geller AM, **Sieving PA.** How many foveal cones are required to "see?": counting photoreceptors in Stargardt's macular dystrophy and modeling with degenerate visual arrays. In: *Retinal Degeneration: Clinical and Laboratory Applications.* (eds.: JG Hollyfield, RE Anderson, MM LaVail). Plenum Publishing Co., New York, 25–34, 1993.

1995

Sieving PA. Diagnostic issues with inherited retinal and macular dystrophies. In: *Seminars in Ophthalmology.* WB Saunders Company. 10:279–94, 1995.

1996

Sieving PA. Questions for "ProVision: preferred responses in ophthalmology, series 2" (ed.: GL Skuta). American Academy of Ophthalmology, 1996.

Sieving PA. Electrical signals of the retina and visual cortex. Chap. 13, 1–15. In: *Duane's Foundations of Clinical Ophthalmology* (eds.: W Tasman, EA Jaeger). J B Lippincott, Philadelphia, 1996.

1998

Sieving PA. Juvenile retinoschisis. Chap. 18. In: *Genetic Diseases of the Eye* (ed: EI Traboulsi). Oxford University Press, New York, NY, 1998.

1999

Sieving PA. Retinitis pigmentosa and related disorders." Chap. 6.10 (550–9). In *Ophthalmology, 3rd ed.* (eds.: M Yanoff, J Duker). Mosby International, London, 2009 (2nd ed., 2004; 1st ed., 1999).

Ayyagari R, Kakuk LE, Toda Y, Coats CL, Bingham EL, Szczesny JJ, Felius J, **Sieving PA**. Blue cone monochromacy: macular degeneration in individuals with cone specific gene loss. Chap. 22 (223–34). In: *Retinal Degenerative Diseases and Experimental Therapy* (eds.: JG Hollyfield, RE Anderson, MM LaVail). Plenum Publishing Corp., New York, NY, 1999.

Bush RA, Sugawara T, Iuvone PM, **Sieving PA**. Melatonin receptor blockers enhance photoreceptor survival and function in light damaged rat retina. In: *Retinal Degenerative Diseases and Experimental Therapy* (eds: JG Hollyfield, RE Anderson, MM LaVail). Plenum Publishing Corp., New York, NY, 1999.

Griesinger IB, **Sieving PA**, Ayyagari R. Autosomal dominant macular degeneration localized to chromosome 6q by linkage analysis. In: *Retinal Degenerative Diseases and Experimental Therapy* (eds: JG Hollyfield, RE Anderson, MM LaVail). Plenum Publishing Corp., New York, NY, 175–182, 1999.

2001

Sieving, PA. Electrophysiologie. In: *Précis d'ophtalmologie* (ed: XD Martin). Editions PAYOT, Lausanne, Switzerland. 2001.

Anderson RE, **Sieving PA**, Maude MB, Naash MI. DHA levels in rod outer segments of transgenic mice expressing G90D rhodopsin mutations. In: *New Insights Into Retinal Degenerative Diseases* (eds: RE Anderson, MM LaVail, JG Hollyfield). Plenum Publishing Corp., New York. 2001.

Breuer DK, Affer M, Andreasson S, Birch DG, Fishman GA, Heckenlively JR, Hiriyanna S, Hoffman DR, Jacobson SG, Mears AJ, Musarella MA, Redolfi B, **Sieving PA**, Wright AF, Yasher BM, Zucchi I, Swaroop A. X-linked retinitis pigmentosa: current status. In: *New Insights Into Retinal Degenerative Diseases* (eds: RE Anderson, MM LaVail, JG Hollyfield). Plenum Publishing Corp., New York. 2001.

Hiriyanna KT, Singh R, Bingham EL, Kemp JA, Ayyagari R, Yashar B, **Sieving PA**. Searching for genotype-phenotype correlations in X-linked juvenile retinoschisis. In: *New Insights Into Retinal Degenerative Diseases* (eds: RE Anderson, MM LaVail, JG Hollyfield). Plenum Publishing Corp., New York. 2001.

2003

Sieving PA, MacDonald IM, Khan NW. Juvenile X-linked retinoschisis. Chap. 79. In: *Handbook of Clinical Electrophysiology of Vision Testing, 2nd Ed* (eds.: J Heckenlively, G Arden). Mosby Year Book Publishers, St. Louis, 2003.

Sieving PA, Meltzer M, MacDonald I. Gene reviews: clinical genetic information resource. June, 2003. GeneReviews (TM) at the GeneTests–GeneClinics Web site. Available at http://www.geneclinics.org, 2003–05.

2004

Sieving PA. Healthy vision month, May 2004: focus is on diabetic retinopathy. *Optometry*. 75:271–3, 2004.

Insel TR, Volkow ND, Landis SC, Li TK, Battey JF, **Sieving PA**. Limits to growth: why neuroscience needs large-scale science. *Nat Neurosci*. 7:426–527, 2004.

2005

Sieving PA, MacDonald IM, Trese MT. Congenital X-linked retinoschisis. Chap. 25, 377–85. In: *Pediatric Retina: Medical and Surgical Approaches* (eds: ME Hartnett, MT Trese, A Capone, et al.). Lippincott Williams & Wilkins, Philadelphia, 2005.

Musarella MA, MacDonald IM, **Sieving, PA**. Retinoschisis, juvenile (X-linked). In: *Encyclopedia of Medical Genomics and Proteomics*. (eds: J Fuchs, M Podda). Marcel Dekker, Inc., New York, 1147–50, 2005.

2006

Sieving P, Macdonald I, Meltzer MR (updated January 2006) X-linked juvenile retinoschisis in: GeneReviews at GeneTests: Medical Genetics Information Resource [database online]. Copyright, University of Washington, Seattle. 1997–2006. Available at http://www.genetests.org.

2008

Vijayasarathy C, Takada Y, Zeng Y, Bush RA, **Sieving PA**. Organization and molecular interactions of retinoschisin in photoreceptors. In *Recent Advances in Retinal Degeneration Research* (eds: RE Anderson, MM LaVail, JG Hollyfield). Springer, New York. NY, 2008, 291–9.

2010

Sieving PA. Foreword to *Ocular Disease: Mechanisms and Management* (eds. LA Levin, DM Albert), WB Saunders (in press).

Sieving PA, Ziccardi L. Retinoschisis. Chap. 18. In: *Genetic Diseases of the Eye, 2nd ed.* (ed: EI Traboulsi). Oxford University Press, New York, NY, (in press).