CURRICULUM VITAE

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William and Mary Jane Brinton Distinguished Professorship in Neurological Diseases

Director
Institute for Neurodegenerative Diseases
UCSF School of Medicine

Professor
Department of Neurology
Department of Biochemistry and Biophysics
UCSF School of Medicine

Professor in Residence
Department of Virology
UC Berkeley School of Public Health

EDUCATION

1960	Walnut Hills High School, Cincinnati, Ohio
1964	A.B. (*cum laude*), University of Pennsylvania, The College, Philadelphia, PA
1968	M.D., University of Pennsylvania, School of Medicine, Philadelphia, PA
1968 – 1969	Internship in Medicine, University of California, School of Medicine, San Francisco, CA
1972 – 1974	Residency in Neurology, University of California, School of Medicine, San Francisco, CA

DIPLOMATE

1979	Certified in Neurology, American Board of Psychiatry and Neurology, Certificate No. 18907

LICENSURE

1969	California – G 022619

APPOINTMENTS

1974 – 1980	Assistant Professor of Neurology in Residence, University of California, School of Medicine, San Francisco, California
1976 – 1988	Lecturer, Department of Biochemistry and Biophysics, University of California, School of Medicine, San Francisco, California
1979 – 1983	Assistant Professor of Virology in Residence, University of California, School of Public Health, Berkeley, California
1980 – 1981	Associate Professor of Neurology in Residence, University of California, School of Medicine, San Francisco, California
1981 – 1984	Associate Professor of Neurology, University of California, School of Medicine, San Francisco, California
1983 – 1984	Associate Professor of Virology in Residence, University of California, School of Public Health, Berkeley, California
1984 – present	Professor of Neurology, University of California, School of Medicine, San Francisco, California

1984 – present	Professor of Virology in Residence, University of California, School of Public Health, Berkeley, California
1994	Visiting Professor, College de France, Paris, France
1988 – present	Professor of Biochemistry, University of California, School of Medicine, San Francisco, California
1999 – present	Director, Institute for Neurodegenerative Diseases, University of California, School of Medicine, San Francisco, California
1999	Visiting Professor, Garvin Institute, Sydney, Australia
2007 – 2008	Leverhulme Visiting Professor, Imperial College, London, United Kingdom

COMMENCEMENT ADDRESSES

2001	Commencement Address, Pennsylvania State University, Schools of Medicine and Graduate Studies, Hershey, Pennsylvania, May 20, 2001
2007	Commencement Address, Keck Graduate Institute of Applied Life Sciences, Claremont, California, May 13, 2007
2010	Commencement Address, Rosalind Franklin University of Medicine and Science, Chicago, Illinois, June 4, 2010

PROFESSIONAL SOCIETIES

American Society of Biochemistry and Molecular Biology
American Society for Clinical Investigation, Emeritus
American Chemical Society
American Association for the Advancement of Science, 1997, Fellow
American Academy of Neurology, 1996, Fellow; 2003, Honorary Member
American Society for Neurochemistry
American Society for Microbiology, 1997, Fellow
International Society for Neurochemistry
American Society for Cell Biology
California Medical Association
San Francisco Neurological Society
Society for Neuroscience
American Neurological Association, 1997, Honorary Member
American Society of Virology
American Society of Human Genetics
Association of American Physicians
Association of California Neurologists
American Association of Anatomists, 2015, Honorary Member
American Society of Neuroradiology, 2016, Honorary Member

OTHER AFFILIATIONS

1974 – 1999	Attending Neurologist, Veterans Administration Hospital, Fort Miley, San Francisco, CA
1974 – present	Attending Neurologist, San Francisco General Hospital, San Francisco, CA
1990 – present	Member, Program in Biological Sciences, Molecular Medicine Section, University of California, San Francisco, CA
1994 – present	Member, Program in Biological Sciences, Neuroscience Section, University of California, San Francisco, CA
2007 – present	Member, Institute for Regenerative Medicine, University of California, San Francisco, CA

HONORS AND AWARDS

1963	Phi Beta Kappa
1963	Alpha Epsilon Delta (premedical honor society)
1963	Sigma Xi
1967	Alpha Omega Alpha
1968	Roy G. Williams Basic Science Research Award, University of Pennsylvania, School of Medicine, Philadelphia, Pennsylvania
1975 – 1976	Research Career Development Award, National Institutes of Health, United States Public Health Service
1976 – 1978	Alfred P. Sloan Foundation Research Fellow, New York, New York
1976 – 1981	Howard Hughes Medical Investigator
1985 – 1990	Senator Jacob Javits Center for Excellence in Neuroscience, National Institutes of Health, Principal Investigator and Director
1987	George Cotzias Award for Outstanding Research in Neurology, American Academy of Neurology
1989 – 1990	33rd Faculty Research Lecturer, University of California San Francisco Academic Senate
1990 – 1997	Outstanding Investigator Award for Leadership and Excellence in Alzheimer's Disease, National Institutes of Health
1991	Potamkin Prize for Alzheimer's Disease Research, American Academy of Neurology
1991	Distinguished Medical Graduate Award, University of Pennsylvania, Philadelphia, Pennsylvania
1992	Christopher Columbus Quincentennial Discovery Award in Biomedical Research, National Institutes of Health and Medical Society of Genoa, Italy
1992	Metropolitan Life Foundation Award for Medical Research, New York, New York
1992	National Academy of Sciences of the United States of America, Member
1992	Institute of Medicine, National Academy of Sciences, Member
1992	Charles A. Dana Award for Pioneering Achievements in Health
1992	Health Science Achievement Award, University of Illinois, Chicago, Illinois
1992	Dickson Prize for Distinguished Scientific Accomplishments in Medicine, University of Pittsburgh
1992	Lifetime Science Achievement Award, George Washington University, Washington, D.C.
1992	Max-Planck-Forschungspreis, Alexander von Humboldt-Stiftung and Max-Planck-Gesellschaft (Shared with Detlev Riesner)
1993	Presidential Award, American Academy of Neurology
1993	Richard Lounsbery Award for Extraordinary Scientific Research in Biology and Medicine, National Academy of Sciences (Shared with Bert Vogelstein)
1993	Gairdner Foundation International Award for Outstanding Achievement in the Field of Medical Science, Toronto, Canada
1993	American Academy of Arts and Sciences, Fellow
1994	Professeur Associé, Collège de France, Paris, France
1994	Bristol-Myers Squibb Award for Distinguished Achievement in Neuroscience Research
1994	Albert Lasker Award for Basic Medical Research, Lasker Foundation, New York, New York
1995	Paul Hoch Award for Excellence in Research, American Psychopathological Association
1995	Paul Ehrlich and Ludwig Darmstaedter Award, Paul Ehrlich Foundation and the Federal Republic of Germany
1995	Royal College of Physicians, London, England, Foreign Fellow
1995	J. Elliott Royer Award for Contributions to Neurology, University of California San Francisco
1996	Caledonian Research Foundation Prize, Royal Society of Edinburgh, Scotland
1996	Wolf Prize in Medicine, Wolf Foundation and the State of Israel
1996	ICN International Prize in Virology, ICN Pharmaceuticals
1996	Pasarow Prize in Neuroscience, Pasarow Foundation, Los Angeles, California
1996	Prix d'Honneur, Louis Vuitton Moet Hennessy, Paris, France (Shared with Fred Cohen)

1996	Golden Plate Award, American Academy of Achievement
1996	Charles-Leopold Mayer Prize, Institut de France, Academie des Sciences, Paris, France (Shared with Charles Weissmann)
1996	Keio International Award for Medical Science, Keio University, Tokyo, Japan
1996	Baxter Award for Distinguished Medical Research, American Assoc. of Medical Colleges
1996	ICAAC Award, American Society of Microbiology
1996	Rehfuss Medal for Distinguished Service to Medicine, Thomas Jefferson University, Philadelphia, Pennsylvania
1997	Theobald Smith Award, Albany Medical College, Albany, New York
1997	Victor and Clara Soriano Award, World Federation of Neurology
1997	Amgen Award, Protein Society
1997	K. J. Zülch Prize for Basic Neurological Research, Gertrud Reemtsma Stiftung, Max-Planck-Gesellschaft (Shared with Charles Weissmann)
1997	Royal Society, London, England, Foreign Member
1997	Louisa Gross Horwitz Prize, Columbia University, New York, New York
1997	Henry N. Neufeld Memorial Award, United States–Israel Binational Science Foundation (Shared with Zeev Meiner)
1997	Nobel Prize in Physiology or Medicine, Nobel Foundation, Stockholm, Sweden
1998	Jubilee Medal, Swedish Medical Society, Stockholm, Sweden
1998	Franklin Institute Gold Medal, Benjamin Franklin Institute, Philadelphia, Pennsylvania
1998	UCSF Medal, University of California, San Francisco
1998	American Philosophical Society, Member
1998	Medalla Recoral, Universidad de Chile, Santiago, Chile
1998	Founding Associates Award, John Douglas French Foundation, Los Angeles, California
1998	Distinguished Achievement Award, American Academy of Neurology
1999	Prize Lecture Medal, University College London, London, England
1999	Sir Hans Krebs Medal, Federation of European Biochemical Societies
1999	International Fellowship, Garvan Institute, Sydney, Australia
1999	Achievement in Science Award, American Committee for Shaare Zedek Medical Center in Jerusalem, San Francisco, California
1999	Académie Royale de Medecine de Belgique, Foreign Member, Brussels, Belgium
1999	American Neurological Association, Honorary Member
2000	Ellen Browning Medal, Scripps Foundation for Medicine and Science, La Jolla, California
2003	Distinguished Alumni Award, University of Pennsylvania, Philadelphia, Pennsylvania
2003	American Academy of Neurology, Honorary Member
2003	World Jewish Academy of Sciences, Honorary Member
2003	Serbian Academy of Sciences, Foreign Member
2004	Commonwealth Award for Distinguished Service in Science, Wilmington, Delaware
2005	William Beaumont Medal, Wayne County Medical Society, Detroit, Michigan
2006	Fourth Military Medical University, Beijing, China, Honorary Professor
2006	Ernst Knobil Award, University of Texas Medical School, Houston, Texas
2007	T. S. Srinivasan Gold Medal, Chennai, India
2007	Leverhulme Trust Fellowship, London, United Kingdom
2008	Académie d'Agriculture de France, Member, Paris, France
2009	Society for General Microbiology Prize Medal, Harrogate, England
2009	George Eastman Medal, The College of the University of Rochester, Rochester, New York
2010	2009 National Medal of Science, Presidential Award, The National Science Foundation, Washington, D.C.
2011	Distinguished Alumni Award in Chemistry, University of Pennsylvania, Philadelphia, Pennsylvania
2015	American Association of Anatomists, Honorary Member
2015	150th Anniversary Alumni Excellence Award, University of California San Francisco, San Francisco, California

2016 ASNR Honorary Member Award, American Society of Neuroradiology, Washington, D.C.
2016 Cozzarelli Prize, National Academy of Sciences, *Proceedings of the National Academy of Sciences of the United States of America*, Washington, D.C.
2016 BrightFocus Award, BrightFocus Foundation, Clarksburg, Maryland
2020 Bishop Dr. Karl Golser Award, Bishop Dr. Karl Golser Foundation, Knight's hall, Trostburg, Waidbruck, Italy

HONORARY DEGREES

1995 Doctor of Philosophy Honoris Causa, Hebrew University, Jerusalem, Israel
1996 Doctor of Philosophy Honoris Causa, Université René Descartes–Paris V, France
1998 Doctor of Science Honoris Causa, University of Pennsylvania, Philadelphia, Pennsylvania
1999 Doctor of Science Honoris Causa, Dartmouth College, Hanover, New Hampshire
2000 Laurea ad Honorem in Medicina e Chirurgia, Dell'Università di Bologna, Bologna, Italy
2000 Docteur Honoris Causa, l'Université de Liège, Liège, Belgium
2001 Doctor of Science Honoris Causa, Pennsylvania State University, Hershey, Pennsylvania
2005 Doctores Honoris Causa, Universidad Cardenal Herrera-CEU, Moncada, Spain
2007 Doctor of Philosophy Honoris Causa, Claremont Colleges, Claremont, California
2010 Doctor of Philosophy Honoris Causa, Rosalind Franklin University, Chicago, Illinois
2015 Doctor of Science Honoris Causa, Mount Sinai School of Medicine, New York, New York

OTHER HONORS

2008 Stanley B. Prusiner Medical Information Center, Hadassah University Hospital, Jerusalem, Israel
2013 The Prusiner-Abramsky Neuroscience Research Awards to young investigators at Hebrew University of Jerusalem

SERVICE AND COMMERCIAL ACTIVITIES

MILITARY SERVICE

1969 – 1972 Research Associate, Lt. Commander, U.S. Public Health Service, National Institutes of Health, National Heart and Lung Institute, Laboratory of Biochemistry, Section on Enzymes, Bethesda, Maryland

UNIVERSITY SERVICE

1979 – 1982 Institutional Biosafety Committee, *Vice-Chairman*
1980 – 1981 Biosafety Manual Subcommittee, *Chairman*
1984 – 1985 Searle Scholar Nominating Committee
1989 *Ad hoc* Committee to Investigate Scientific Misconduct, *Chairman*
1989 – 1991 Nuclear Magnetic Resonance Scientific Advisory Committee
1989 – 1991 Planning Committee for a Second UCSF Campus
1989 – 1996 Committee on Animal Research
1991 – 1997 Fellowship Selection Committee, Program in Biological Science, Molecular Medicine Section
1992 – 1994 Academic Planning Committee, Letterman/LAIR
1993 Cell Culture Facility Advisory Committee
1993 *Ad hoc* Psychiatry Department Review Committee, *Chairman*
1994 – 1995 Committee on Indirect Costs
1994 – 1996 Chancellor's Task Force for Development of New Initiatives at SFGH
1994 – 1997 UCSF Medal Award Selection Committee
1995 *Ad hoc* Radiobiology Search Committee for Directorship
1997 Chancellor's Committee for Development of the Mission Bay Campus
1997 – 2000 Advisory Committee, Institute for Health and Aging Policy, School of Nursing
2001 Faculty Member, Western Oral Research Consortium
2009 Neuroscience Initiative Committee, *Member*
2010 – present Sandler Neurosciences Center, Building Committee, *Member*
2010 – 2012 Research Strategy Workgroup, UC Commission on the Future, *Member*
2010 – 2012 Academic Council, Special Committee on the Future of the University, *Member*
2016 – present Weill Institute for Neurosciences, Executive Committee, Member
2016 – 2017 UCSF Department of Neurology, Chair Search Committee, Member

MEDICAL SCHOOL SERVICE

1977 – 1986 University of California Santa Cruz-University of California San Francisco Minority Student Training Program Committee
1979 – present *Ad hoc* Promotion Committees
1980 – 1985 Medical Student Research Program Committee
1981 Medical Student Screening Committee
1985 Psychiatry Chairman Search Committee
1985 Dean's Scientific Colloquia Committee
1989 – 1991 Psychiatry Search Committee for Alzheimer's Disease Research
1989 – 1995 Member, Scientific Advisory Board, Ernest Gallo Clinic and Research Center
1994 Working Group for a Department of Human Genetics
1995 Search Committee for Director of Human Genetics Program
1995 Search Committee for Director of Genetics Division, Department of Medicine
1996 – 1997 Planning Committee for Mission Bay Campus
1996 – 2014 Scientific Advisory Board, Ernest Gallo Clinic and Research Center, *Chairman*
2000 – 2002 Dean's Executive Committee, School of Medicine, *Member*
2001 – 2005 Physician-Scientist Research Program Committee, *Member*
2001 – 2005 Advisory Board, ALS Center, *Member*
2011 – 2012 Psychiatry Chairman Search Committee

INSTITUTE FOR NEURODEGENERATIVE DISEASES SERVICE

1999 – present	Founding Director
1999 – present	Executive Committee, Chairman
2013 – present	Ad hoc Search Committees
2012 – 2014	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Chemical Biology
2013 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Molecular/Cell Biology
2013 – 2015	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Computational Biology
2013 – 2015	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Neuropathology
2013 – 2015	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Neurogenetics
2015 – 2018	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Computational Chemistry
2015 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Medicinal Chemistry
2015 – 2018	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Nuclear Chemistry/Radiology
2015 – 2018	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Pharmacokinetics/Neuropharmacology
2015 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, High-Throughput Screening
2016 – 2018	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Neuropharmacology
2017 – 2020	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Alzheimer's Disease Neurobiology
2017 – 2018	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Nuclear Positron Emission Tomography (PET)
2019 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Mass Spectrometry Imaging/Proteomics
2019 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Neurobiology
2019 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Neuropathology
2019 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Chemistry Specialist
2019 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Structural Biology
2019 – present	Ad hoc Search Committee, Institute for Neurodegenerative Diseases, Cryo-Em Structural Biology

DEPARTMENTAL SERVICE

1975 – 1982	Neurology Research Seminar Coordinator
1981 – 1982	Residency Neuroscience Conference Director
1981 – 1984	Search Committee for Faculty Appointments at Veterans Administration Hospital in Neurology
1985 – 1987	Joint Neurobiology-Neurology Seminar Series Committee
1989	Committee to Develop a Scientific Writing Course in Biochemistry, *Chairman*
1993 – 1995	Decade of the Brain Fund Raising Committee, *Chairman*
1993 – present	Neurology Executive Committee, *Member*
1996 – 1997	Biochemistry and Biophysics Committee for Gordon Tompkins Lecture Program
1998 – 2005	Executive Committee, Sandler Neurogenetics Center, *Member*

EDITORIAL SERVICE

1976 – present	*Ad hoc referee:* Biochemistry, Archives of Biochemistry and Biophysics, Journal of General Virology, Canadian Journal of Biochemistry, Western Journal of Medicine, Science, Nature, Journal of Virology, Intervirology, Archives of Virology, American Journal of Pathology, Annals of Neurology, Molecular and Cellular Biology, New England Journal of Medicine, Journal of Infectious Diseases, Nucleic Acids Research, Experimental Neurolog, European Molecular Biology Organization (EMBO)
1979 – 1985	Journal of Neurochemistry, *Editorial Board*

1987 – 1993 Neurology, *Editorial Board*
1990 – 1999 Brain Pathology, *Editorial Board*
1990 – 2010 Reviews in Medical Virology, *Editorial Board*
1991 – 1996 Glycobiology, *Editorial Board*
1993 – present Neurobiology of Disease, *Editorial Board*
1993 – 2002 Amyloid: International Journal of Experimental & Clinical Investigation, *Editorial Board*
1994 – 2004 Molecular Medicine, *Editorial Board*
1995 – 1998 Current Biology – Folding and Design, *Editorial Board*
1995 – 1999 Proceedings of the American Association of Physicians, *Editorial Board*
1995 – 2005 Laboratory Investigations, *Editorial Board*
1996 – 2008 Proceedings of the National Academy of Sciences USA, *Editorial Board*
1996 – 1999 FASEB Journal, *Editorial Board*
1997 – 2016 JAMA Neurology, *Editorial Board*
2000 – present Clinical Neuroscience Research, *Editorial Board*
2002 – present Upper Board, Wiley-VCH Encyclopedia, The Encyclopedia of Molecular Biology and Molecular Medicine
2003 – present Discovery Medicine, *Senior Editor*
2006 – present Prion, *Editorial Board*
2008 – 2011 The American Journal of Pathology, *Reviews Editor*

INTERNATIONAL SERVICE

1998 – 2002 Peres Center for Peace, Tel Aviv, Israel, *Medical Science Advisor*
1999 – 2014 Agnes Ginges Center for Neurogenetics, Department of Neurology, Hadassah Medical School and Hospital, Hebrew University, Jerusalem, Israel, *Executive Committee Chairman*
2001 – 2002 Institute of Catalysis Science and Technology, Technion – Israel Institute of Technology, Haifa, Israel, *Scientific Advisory Board, Member*
2002 – 2005 Board of Directors, La Foundation Ipsen, Paris, France, *Board of Directors, Member*
2003 – 2006 Cambridge Centre for Stem Cell Biology and Medicine, University of Cambridge, Cambridge, United Kingdom, *International Scientific Advisory Board, Member*
2015 Prion 2015 International Congress, *Scientific Advisory Board, Member*

NATIONAL SERVICE

1976 – 2012 National Science Foundation research grants, *Ad hoc referee*
1978 National Institutes of Health, National Institute of Neurological and Communicative Disorders and Stroke, National Research Strategy Panel on Inflammatory, Demyelinating, and Degenerative Diseases, *Consultant*
1981 – 1985 Scientific Program Committee, American Academy of Neurology
1981 – 2015 National Institutes of Health research grants, *Ad hoc referee*
1982 – 1986 Neurological Disorders Program – Project Review A Committee, National Institutes of Health
1984 – 1995 John D. French Foundation for Alzheimer's Disease, Los Angeles, California, Scientific Advisory Board, *Member*
1985 Alzheimer's Disease and Related Disorders Association research grants, *Ad hoc referee*
1985 – 1986 United States Department of Agriculture research grants, *Ad hoc referee*
1986 National Institutes of Health, Division of Research Grants, Special Review Committee*, Chairman*
1986 – 1994 American Health Assistance Foundation, Rockville, Maryland, Scientific Advisory Board
1988 & 1992 National Institutes of Health, National Institute of Aging, Panel for Leadership and Excellence Awards in Alzheimer's Disease, *Ad hoc referee*
1989 National Institutes of Health, National Institute of Aging, Committee for Alzheimer's Disease Research Centers, *Ad hoc referee*
1989 – 1994 American Health Assistance Foundation, Rockville, Maryland, Scientific Advisory Board*, Chairman*

1990 – 1992	National Institutes of Health, National Institute of Neurological Disorders and Stroke, Training Grant and Career Development Review Committee
1990 – 1992	American Academy of Neurology, Fundraising Task Force
1990 – 1994	American Neurologic Association, Long Range Planning Committee
1991 – 1996	American Academy of Neurology Foundation for Education and Research, *Trustee*
1992	Neurodegenerative Diseases Research Center, University of Toronto, Toronto, Canada, *Review Committee*
1994	Alzheimer's Disease Research Planning Meeting, Washington, D.C.
1994 – 2001	Potamkin Prize in Pick's and Alzheimer's Diseases Research Selection Committee, American Academy of Neurology
1995	National Academy of Sciences, Nominating Committee, *Member*
1995 – 1997	National Academy of Sciences, Section 41, Search and Screening Committee
1995 – 1997	American Neurological Association, *Councilor*
1995 – 2000	American Health Assistance Foundation, Rockville, Maryland, Scientific Advisory Board, *Special Advisor*
1996 – 2013	John D. French Foundation for Alzheimer's Disease, Los Angeles, California, Scientific Advisory Board, *Chairman*
1997 – 2001	Center for Biologics Evaluation and Research, Food and Drug Administration, Transmissible Spongiform Encephalopathies Advisory Committee, *Member*
1998	National Academy of Sciences, Richard Lounsbery Award Committee, *Chairman*
1998 – 2001	Bard College, Annandale-on-Hudson, New York, Committee to Develop a New Science Curriculum, *Chairman*
1998 – 2002	Institute of Medicine, Chair Section Liaison
1999 – 2002	Institute of Medicine, Membership Committee
1999 – 2002	University of Texas Southwestern Medical Center, Dallas, Texas, External Advisory Committee for Endowed Program for Scholars in Biomedical Research, *Member*
1999 – 2004	American Academy of Achievement, Washington, D.C., Awards Council, *Member*
2000 – 2005	University of Pennsylvania, Philadelphia, Pennsylvania, *Trustee*
2000 – 2004	National Advisory Council on Aging, National Institutes of Health, Bethesda, Maryland, *Member*
2001 – 2004	Center for Biologics Evaluation and Research, Food and Drug Administration, *Consultant*
2002 – 2004	Department of Defense National Prion Research Program, Institute of Medicine Study Panel, *Consultant*
2003 – 2005	American Association for the Advancement of Science, Committee on Nominations, *Member*
2002 – 2016	University of Texas Southwestern Medical Center, Dallas, Texas, External Advisory Committee for Endowed Program for Scholars in Biomedical Research, *Chairman*
2002 – 2014	Foundation for Biomedical Research, Washington, D.C., Board of Governors, *Member*
2003 – 2005	American Association for the Advancement of Science, Committee on Nominations, *Member*
2003 – 2014	Honorary Board of the International Raoul Wallenberg Foundation and the Angelo Roncalli International Committee, New York, NY, *Member*
2003 – 2010	International Longevity Center – USA, Board of Directors, *Member*
2007 – 2010	National Academy of Sciences Council, *Elected Member*
2007 – 2010	Budget and Internal Affairs Committee, National Academy of Sciences Council, *Member*
2007 – 2010	Executive Compensation Committee, National Academy of Sciences Council, *Member*
2007 – 2013	National Society of High School Scholars, Atlanta, Georgia, *Board of Directors*
2008	Committee on Improving the Organization of the US Department of Health and Human Services to Advance the Health of Our Population, Institute of Medicine, *Member*
2008 – 2009	Governing Board, National Research Council, Washington, D.C., *Member*
2008 – 2009	Institute of Medicine Study Panel, "Organizing the Department of Health and Human Services to Achieve a Healthier America", *Consultant*
2008 – 2010	Scientific Program Committee, National Academy of Sciences Council, *Member*
2008 – 2011	National Research Council Governing Board, National Academy of Sciences, *Member*
2008 – 2015	American Health Assistance Foundation, Clarksburg, Maryland, *Board of Directors*

2008 – 2012 Science and Entertainment Exchange Advisory Board, National Academy of Science, *Member*
2009 – 2010 National Academy of Sciences, Washington, D.C., Executive Committee, *Member*
2009 – 2010 National Academy of Sciences, Washington, D.C., Executive Compensation Committee, *Chairman*
2010 – 2011 National Research Council, National Academy of Sciences, Committee on Future Science Opportunities in the Antarctic and Southern Ocean, *Member*
2010 – 2012 National Academy of Sciences, Washington, D.C., Building Restoration Committee, *Member*
2012 – present USAgainstAlzheimer's Network (USA2), Washington, D.C., Board of Directors, *Member*
2012 – 2017 Cleveland Clinic Lou Ruvo Center for Brain Health, Scientific Advisory Board, *Chairman*
2013 – present Blavatnik Awards for Young Scientists, Scientific Advisory Board, *Member*
2013 – 2015 American Neurological Association, *President Elect*
2014 – present Weill Cornell Medical College, New York, New York, Board of Fellows, *Member*
2014 – present Foundation for Food and Agriculture Research, Washington, D.C., *Member*

COMMUNITY SERVICE

1982 – 1990 Advisory Board, Family Survival Project for Adults with Chronic Brain Disorders
1985 – 1990 Scientific Review Committee, State of California Alzheimer's Disease Diagnostic Center and Research Grant Program
1985 – 1992 Advisory Board, Alzheimer's Disease and Related Disorders Association, San Francisco Chapter
1989 Chairman, Scientific review Committee, State of California Alzheimer's Disease Diagnostic Center and Research Grant Program
1995 – 2005 Board of Directors, Concordia Argonaut Club, San Francisco, California
1999 – 2002 Board of Trustees, Sherith Israel Synagogue, San Francisco, California
2002 – 2007 Board of Directors, Fromm Institute for Lifelong Learning, San Francisco, California

COMMERCIAL ACTIVITIES

1996 – 1999 Centeon LLP, Transmissible Agent Safety Committee (TASC), *Member*
1998 – 1999 United Airlines Inc., Advisory Committee for Improving Airplane Air Quality, *Chairman*
1999 – 2005 KBC Pharma Inc., Board of Directors, *Member*
2001 – 2008 InPro Biotechnology, Inc., Board of Directors, *Chairman*
2014 – 2016 Emmaus Medical, Inc., Scientific Advisory Board, *Member*
2016 BioBlast Inc., Scientific Advisory Board, *Chairman*
2016 – 2019 Alzheon Inc., Scientific Advisory Board, *Chair*
2018 – present Trizell Inc., Board of Directors, *Member*
2018 – present ViewPoint Therapeutics Inc., Scientific Advisory Board, *Member*
2018 – present New Ventures Inc., Scientific Advisory Board, *Member*

HONORARY LECTURES

Honorary Lectures

1) Ray A. and Robert C. Kroc Lecture, University of California Los Angeles, Department of Neurology, Los Angeles, California,
September 26, 1985,
"The Structure and Biology of Prions Causing Dementia."

2) Andrew Mark Lippard Memorial Lecture, Columbia University, College of Physicians and Surgeons, M.D Ph.D. Program and Department of Neurology, New York, New York,
October 8, 1985,
"Prions Causing Scrapie and Creutzfeldt-Jakob Disease."

3) Louis B. Flexer Lecture, Neurosciences Institute, University of Pennsylvania, Philadelphia, Pennsylvania,
May 6, 1986,
"Prions — Novel Infectious Pathogens Causing Brain Degeneration."

4) Dean's Distinguished Professor Lecture, Eastern Virginia Medical School, Norfolk, Virginia,
September 30, 1986,
"Investigations into the Process of Aging."

5) Grass Foundation Traveling Scientist Lecture, Louisiana State University, Baton Rouge Chapter of the Society of Neuroscience, Baton Rouge, Louisiana,
November 4, 1986,
"Prions, Brain Degeneration, and Alzheimer's Disease."

6) Cotzias Award Lecture, American Academy of Neurology Meeting, New York, New York,
April 5, 1987,
"Prions — Novel Infectious Pathogens Causing Scrapie and Creutzfeldt-Jakob Disease."

7) Kenneth M. Campione Visiting Professor of Neurology, Northwestern University, Chicago, Illinois,
March 8, 1988,
"Prions in the Central Nervous System Diseases."

8) S. Stanley Schneierson Memorial Lecture, Mt. Sinai School of Medicine of the City University of New York, Departments of Microbiology and Medicine and the Page and William Black Post-Graduate School of Medicine, New York, New York,
May 3, 1989,
"Molecular Biology and Structure of Prions Causing Infectious, Sporadic, and Genetic Diseases of Humans."

9) Dean's Distinguished Seminar Speaker, University of Colorado, Denver, Colorado,
October 4, 1989,
"Unraveling Prion Diseases through Molecular Genetics and Transgenic Mice."

10) Faculty Research Lecture, Academic Senate, University of California San Francisco, San Francisco, California,
May 9, 1990,
"Unraveling Prion Diseases through Molecular Biology and Transgenetics."

11) Potamkin Prize for Alzheimer's Disease Research, 43rd Annual Meeting of the American Academy of Neurology, Boston, Massachusetts,

April 23, 1991,
"Molecular Biology and Transgenetics of Prion Diseases: A Renaissance in Studies of Neurodegenerative Disorders in Humans and Animals."

12) Distinguished Medical Graduate Award, University of Pennsylvania, School of Medicine, Philadelphia, Pennsylvania,
May 17, 1991,
"Molecular Biology and Transgenetics of Prion Diseases: A Renaissance in Studies of Neurodegenerative Disorders in Humans and Animals."

13) Janssen Presidential Lecture, XVth International Symposium on Cerebral Blood Flow and Metabolism, International Society for Cerebral Blood Flow and Metabolism, Miami, Florida,
June 4, 1991,
"CNS Degeneration Caused by Prions — Genetic and Infectious Mechanisms."

14) Gesellschaft Föderung der Molekularbiologischen Forschung Lecture, Zentrum für Molekulare Biologie, ZMBH Forum: Cells and Signaling, Heidelberg, Germany,
September 30, 1991,
"Unraveling Prions through Molecular Biology and Transgenetics."

15) Lilly Lecture, Lilly Research Laboratories, Indianapolis, Indiana,
October 4, 1991,
"Prion Diseases of Humans and Animals."

16) Mary and Sandy McEwan Memorial Neurology Lecture, University of Toronto, Division of Neurology, Toronto, Ontario, Canada,
October 24, 1991,
"Prion Biology — Infectious and Genetic Mechanisms in CNS Degeneration."

17) Robert R. Kohn Memorial Lecture, Case Western Reserve University, School of Medicine, Cleveland, Ohio,
October 31, 1991,
"The Molecular Biology and Chemistry of Prions."

18) Harry M. Rose Lecture on Infectious Diseases, Columbia College of Physicians and Surgeons, New York, New York,
November 13, 1991,
"Infectious and Genetic Prion Diseases of Humans and Animals."

19) Melville A. Hare Memorial Lecture, University of Rochester, School of Medicine and Dentistry, Microbiology and Immunology Department Seminar, Rochester, New York,
December 11, 1991,
"Prion Diseases of Humans and Animals — Genetic and Infectious Mechanisms."

20) Harvey Lecture, The Harvey Society, Rockefeller University, New York, New York,
March 26, 1992,
"Prion Diseases."

21) Gordon Holmes Memorial Lecture, Third Meeting of the European Neurological Society, Lausanne, Switzerland,
June 29, 1992,
"Prion Encephalopathy."

22) Merck Centennial Lecture, University of Kansas, School of Pharmacy, Lawrence, Kansas, September 24, 1992,
"Molecular Biology of Prion Diseases in Humans and Animals."

23) Fondazione Sigma-Tau, Lectures on Aging, Rome, Italy,
October 12, 1992,
"Biologica Moleculare e Genetica delle malattie da Prioni: Degenerazione del SNC in relazione all'ettà."

24) Dickson Prize in Medicine, University of Pittsburgh, Pittsburgh, Pennsylvania,
December 10, 1992,
"Degenerative Brain Diseases Caused by Prions."

25) Lounsbery Award Lecture, National Academy of Sciences One Hundred and Thirtieth Annual Meeting, Washington, D.C.,
April 25, 1993,
"Prions in Biology and Medicine."

26) The David Oppenheimer Memorial Lecture, University of Oxford, Neuroscience Lecture Series, Department of Neuropathology, Oxford, England,
September 24, 1993,
"Chemistry, Biology and Genetics of Prion Diseases."

27) Gairdner International Award Lecture, University of Toronto, Toronto, Canada,
October 21, 1993,
"Prions and Brain Degeneration."

28) Gairdner International Award Lecture, McGill University, Montreal General Hospital Research Institute, Centre for Research in Neuroscience, Montreal, Canada,
October 25, 1993,
"Molecular Biology and Genetics of Prion Diseases."

29) Lectureship Award on Basic Cell Research in Cytology, American Society of Cytology, Houston, Texas,
November 4, 1993,
"Prions Causing CNS Degeneration in Humans and Animals."

30) Carl Vernon Moore Memorial Lecture, Washington University, Department of Medicine, St. Louis, Missouri,
November 5, 1993,
"Transgenetic and Protein Structural Approaches to Unraveling the Enigmas of Prion Diseases."

31) Seventh Annual Willard B. Rew Memorial Lecture, Harvard Medical School, Boston, Massachusetts,
May 3, 1994,
"Age Dependent Dementias and Neurodegeneration Caused by Prions."

32) Anders Retzius Lecture, Karolinska Institutet, Stockholm, Sweden,
May 17, 1994,
"Molecular Biology and Genetics of Prion Diseases."

33) Sackler Visiting Professor and Lecture, Cornell University School of Medicine, New York Hospital, Department of Neurology, New York, New York,
June 22, 1994,
"Inherited, Infectious, and Sporadic Human Prion Diseases."

34) First Annual Stanley Fahn Lecture, 3rd International Congress of Movement Disorders, Orlando, Florida, November 7, 1994,
"Prion Diseases."

35) Sixth Annual Leonard Tevelson Memorial Lecture, Department of Microbiology and Immunology, Temple University School of Medicine, Philadelphia, Pennsylvania,
November 8, 1994,
"The Biochemistry and Genetics of Prion Diseases."

36) Harry DeLozier Lecture, Department of Neurology, Ohio State University, Columbus, Ohio,
November 17, 1994,
"Clinical Aspects of Prion Disease."

37) Norman Allen Lecture, Department of Neurology, Ohio State University, Columbus, Ohio,
November 17, 1994,
"New Approaches to Neurodegeneration through Studies of Prion Diseases."

38) Perspectives in Comparative Medicine Distinguished Lecturer, Yale University School of Medicine, New Haven, Connecticut,
December 7, 1994,
"Chimeric Prion Protein Transgenes and Molecular Biological Investigations of Prion Diseases."

39) Special Dean's Research Seminar Series Lecture, University of California San Francisco, San Francisco, California,
January 27, 1995,
"Prion Odyssey From Laughing Cannibals to Mad Cows."

40) Distinguished Lecturer in the Life Sciences, Boyce Thompson Institute, Cornell University, Ithaca, New York,
February 15, 1995,
"Molecular, Biological, and Biophysical Investigations of Prions Causing CNS Degeneration."

41) Evans Lecturer, Department of Medicine, Boston University Medical Center, Boston, Massachusetts,
February 16, 1995,
"The Prion Diseases."

42) Hoch Award Lecture, American Psychopathological Association, New York, New York,
March 2, 1995,
"Genetic and Infectious Prion Diseases of Humans and Animals."

43) Paul Ehrlich and Ludwig Darmstaedter Award Ceremony, Paulskirche, Frankfurt, Germany,
March 14, 1995,
"Prion Biology and Diseases: Laughing Cannibals, Mad Cows, and Scientific Heresy."

44) Paul Ehrlich Lecture, Paul-Ehrlich-Institut, Langen, Germany,
March 15, 1995,
"The Prion Diseases: Laughing Cannibals, Mad Cows, and Scientific Heresy."

45) Paul Ehrlich Lecture, National Institutes of Health, Foundation for Advanced Studies, Bethesda, Maryland
April 26, 1995,
"The Present State of Prions."

46) Susan Swerling Lecture, Dana-Farber Cancer Institute, Harvard Medical School, Boston, Massachusetts, May 22, 1995,
"Molecular Biology and Biophysics of Prions Causing CNS Degeneration."

47) Georg Hübscher Memorial Lecture, University of Nottingham, Nottingham, England,
May 24, 1995,
"Molecular Biological and Biophysical Studies of Prions Causing Neurodegeneration."

48) Julian S. Davis Lecture, Mt. Zion Medical Center, University of California San Francisco, San Francisco, California,
June 9, 1995,
"Degenerative Illnesses of the Brain — A Lesson from Prion Diseases."

49) Abraham Flexner Lectures, Vanderbilt University School of Medicine, Nashville, Tennessee,
September 26, 1995,
"Molecular Biology and Genetics of Prion Diseases."
September 27, 1995,
"Prion Diseases of Humans and Animals — Clinical and Neuropathological Aspects."
September 27, 1995,
"How Do Prions Replicate? Lessons from Biophysical Studies of the Prion Protein."

50) Grass Foundation Lecture, University of North Carolina School of Medicine, Chapel Hill, North Carolina,
October 2, 1995,
"Neurobiology and Genetics of Prion Diseases."

51) Osamu Hayaishi Lecture, Kyoto University, Kyoto, Japan,
October 30, 1995,
"The Genetic and Physical Basis of Prion Diseases."

52) Distinguished Lecturer, University of Kentucky, Sanders-Brown Center on Aging, Lexington, Kentucky,
December 12, 1995,
"Prion Disease and Neurodegeneration."

53) William Gies Distinguished Lecture, International Association for Dental Research, San Francisco, California,
March 14, 1996,
"Prion Biology and Diseases."

54) Wolf Prize Lecture, Technion, Israel Institute of Technology, Haifa, Israel,
March 28, 1996,
"Prion Biology and Diseases."

55) Wolf Prize Lecture, Weizmann Institute of Science, Rehovot, Israel,
March 28, 1996,
"Prion Biology and Diseases."

56) Eleventh Annual Pfizer Lecture in Honor of Konrad Bloch, Harvard University, Cambridge, Massachusetts,
April 3, 1996,
"Prion Biology and Diseases: Transgenetic and Biophysical Investigations."

57) Morris B. Bender Memorial Lecture, Mt. Sinai Hospital and School of Medicine, New York, New York, May 1, 1996,
"Disorders of Protein Conformation — Prions in the Mad Cow Crisis."

58) Caledonian Research Foundation Prize Lecture, Royal Society of Edinburgh, Edinburgh, Scotland, May 13, 1996,
"The Molecular Biology and Pathogenesis of Prion Disease: an Agent of Neurodegeneration."

59) William Dick Lecture, The Royal Dick School of Veterinary Studies, University of Edinburgh, Edinburgh, Scotland,
May 15, 1996,
"Prions Causing BSE and Scrapie."

60) Pasarow Prize Lecture, University of Southern California, Los Angeles, California,
May 20, 1996,
"Prions, CNS Degeneration, and Mad Cows."

61) ICAAC Award Lecture, American Society for Microbiology, New Orleans, Louisiana,
September 16, 1996,
"The Prion Odyssey — From Heresy to Orthodoxy."

62) John H. Erskine Lecture in Infectious Diseases, St. Jude Children's Research Hospital, Memphis, Tennessee,
September 17, 1996,
"The Challenge of Learning How Prions Destroy the Brains of Humans and Animals."

63) Associated Universities Incorporated Distinguished Lecture, Brookhaven National Laboratory, Upton, New York,
October 1, 1996,
"Prion Biology and Diseases."

64) John Enders Lecture, Children's Hospital, Harvard University, Cambridge, Massachusetts,
October 2, 1996,
"The Prion Saga — A Journey from Heresy to Orthodoxy."

65) Nobel Forum Lecture, Karolinska Institutet, Stockholm, Sweden,
October 1, 1996,
"Prion Biology and Diseases — The Mad Cow Crisis."

66) Huygens Lecture, Netherlands Organization for Scientific Research, The Hague, The Netherlands,
November 6, 1996,
"Mad Cows, Cannibals and Prions."

67) Oretta Bartolomei Corsi Lecture, University of Florence, Florence, Italy,
November 7, 1996,
"Mad Cows, Creutzfeldt-Jakob Disease, and the Biology of Prions."

68) Martin E. Rehfuss Lecture, Thomas Jefferson University, Philadelphia, Pennsylvania,
November 20, 1996,
"Prion Diseases — Mad Cows and Europeans."

69) Charles-Leopold Mayer Prize Lecture, Institut de France, Academie des Sciences, Paris, France, December 10, 1996,
"The Prion Odyssey — From Heresy to Orthodoxy."

70) Université René Descartes Conference, Faculté des Sciences Pharmaceutiques et Biologiques, Paris, France,
December 11, 1996,
"Prion Odyssey: From Laughing Cannibals to Mad Cows."

71) Keio International Medical Science Prize Lecture, Keio University, Tokyo, Japan,
December 19, 1996,
"Prion Biology and Diseases — Studies on the Structural Diversity of Prion Proteins."

72) Theobald Smith Award Lecture, Albany Medical College, Albany, New York,
February 11, 1997,
"Turmoil in Europe: Mad Cows and Prions."

73) Kosuge Memorial Lectureship, University of California, Davis, California,
April 10, 1997,
Technical Seminar:
"Structural Biology and Genetics of Prions: An Odyssey from Heresy to Orthodoxy."
Student Seminar:
"Laughing Cannibals, Mad Cows, and Scientific Heresy."

74) Werner Heisenberg Lecture, Carl Friedrich von Siemens Foundation, Munich, Germany,
June 3, 1997,
"The Biology, Genetics, and Biophysics of Prions."

75) Maud L. Menten Lecture, University of Pittsburgh, Pittsburgh, Pennsylvania,
June 12, 1997,
"Prions — A Scientific Odyssey from Heresy to Orthodoxy."

76) Amgen Lecture, Eleventh Symposium of the Protein Society, Boston, Massachusetts,
July 13, 1997,
"Structural Biology and Genetics of Prions."

77) Victor and Clara Soriano Award Lecture, XVI World Congress of Neurology, Buenos Aires, Argentina,
September 17, 1997,
"Prions and Neurological Disorders."

78) Zülch Award Presentation, Gertrud Reemtsma Foundation, Köln, Germany,
September 26, 1997,
"Prions and Mad Cows — Embracing Fatal Conformations of Proteins during a Journey from Heresy to Orthodoxy."

79) Eagleson Lecture, American Biological Safety Association Conference, San Diego, California,
October 20, 1997,
"Mad Cows, Demented People, and Biocontainment."

80) Maynard Dewey Lecture, SUNY Stony Brook, Stony Brook, New York,
October 22, 1997,
"Biophysical and Genetic Studies of Prions Causing Neurodegeneration in Humans and Animals."

81) Louisa Gross Horwitz Prize Lecture, Columbia University, New York, New York,
October 23, 1997,
"Prions — Fatal Conformational Changes in Humans and Cows."

82) Charles Yanofsky Award Fund Lecture, Stanford University, Stanford, California,
November 24, 1997,
"Prions — Fatal Conformational Changes in Humans and Cows."

83) Nobel Prize Lecture, Karolinska Institutet, Stockholm, Sweden,
December 8, 1997,
"Prions."

84) Nobel Prize Lecture, University of Uppsala, Uppsala, Sweden,
December 13, 1997,
"Prions."

85) Swedish Medical Society Jubilee Gold Medal Lecture, University of Lund, Lund, Sweden
January 20, 1998,
"Prions."

86) Swedish Medical Society Jubilee Gold Medal Lecture, University of Umea, Umea, Sweden
January 21, 1998,
"Prions."

87) Swedish Medical Society Jubilee Gold Medal Lecture, University of Linskoping, Linskoping, Sweden
January 22, 1998,
"Prions."

88) Swedish Medical Society Jubilee Gold Medal Lecture, University of Göteborg, Göteborg, Sweden
January 23, 1998,
"Prions."

89) Dean's Distinguished Seminar Series, University of Colorado Health Sciences Center, Denver, Colorado,
March 2, 1998,
"Lessons from Prion Biology about Mad Cows and Demented People."

90) 33rd Annual George Gamow Memorial Lecture, University of Colorado, Boulder, Colorado,
March 3, 1998,
"Prions — A New Paradigm in Biology and Medicine."

91) Beach Family Lectureship in Biochemistry, Department of Biochemistry, Purdue University,
West Lafayette, Indiana,
March 24, 1998,
"Prions: From Scorn and Ridicule to Notoriety and Preeminence."

92) University of Pennsylvania School of Medicine Graduation, Philadelphia, Pennsylvania,
May 17, 1998,
"Distinguished Alumnus Address."

93) Capital Science Lecture, Carnegie Institution, Washington, D.C.,
October 13, 1998,
"Prion Biology and Diseases."

94) Florence Mahoney Lecture, National Institutes of Health, National Institute on Aging, Bethesda, Maryland,
October 14, 1998,
"Prion Biology and Diseases — A Saga of Skeptical Scientists, Mad Cows, and Laughing Cannibals."

95) Inaugural Jerome Joseph Landa Lecture, University of Utah, Department of Neurology, Salt Lake City, Utah,
January 11, 1999,
"A Saga of Scientific Discovery — Study of Prions Causing Mad Cow Disease and Fatal Brain Degeneration in Humans."

96) Harrington and Fogan Lecture, University at Buffalo School of Medicine and Biomedical Sciences, Buffalo, New York,
March 22, 1999,
"Clinical and Experimental Neurology of Prion Disease."

97) Seventh Orten Lecture, James M. Orten Memorial Fund, Wayne State University School of Medicine, Detroit, Michigan,
March 23, 1999,
"The Saga of Prion Diseases."

98) 44th George H. Bishop Lecture in Experimental Neurology, Washington University School of Medicine, St. Louis, Missouri,
March 24, 1999,
"The Biology and Genetics of Dementing Diseases Caused by Prions."

99) Prize Lecture, University College London, London, England,
June 17, 1999,
"Prions, Mad Cows, and Creutzfeldt-Jakob Disease."

100) The Sir Hans Krebs Lecture, Federation of European Biochemical Societies, Nice, France,
June 19, 1999,
"Prion Biology and Diseases."

101) Inaugural Keynote Lecture, Buck Center for Research in Aging, Novato, California,
September 30, 1999,
"Looking Forward to Neurodegeneration as We Grow Older."

102) Albert Einstein Memorial Lecture, Princeton Chamber of Commerce, Princeton, New Jersey,
October 6, 1999,
"Prions Devouring the Brains of Mad Cows and Laughing Cannibals."

103) F. E. Bennett Lecture, American Neurological Association, Seattle, Washington,
October 12, 1999,
"Clinical and Basic Investigations of Prion Diseases."

104) Benjamin Lieberman Memorial Lecture, UCSF–Mount Zion Center on Aging, University of California, San Francisco, California,
October 26, 1999,
"Aging and Neurodegeneration."

105) Fred Plum Lecture, University of Washington, Department of Neurological Surgery, Seattle, Washington, January 5, 2000,
"New Approaches to Dementing Illnesses —The Saga of Prions from Heresy to Orthodoxy."

106) Institute of Medicine Distinguished Lecture, University of Iowa, Iowa City, Iowa,
April 6, 2000,
"Prions and the Brain."

107) Shattuck Lecture, Massachusetts Medical Society, Boston, Massachusetts,
May 20, 2000,
"Lessons from Prion Diseases for the More Common Neurodegenerative Disorders."

108) Bass Award Lecture, The Society of Neurological Surgeons, Pasadena, California,
May 22, 2000,
"New Approaches to Dementing Illnesses — the Saga of Prions: From Heresy to Orthodoxy."

109) Osamu Hayaishi Lecture, 18th International Congress of Biochemistry and Molecular Biology, Birmingham, England,
July 19, 2000,
"Molecular Biology and Genetics of Prions."

110) St. Geme Lecture, Department of Neurology, University of Colorado Health Sciences Center, Denver, Colorado,
December 14, 2000,
"Demented People and Mad Cows Caused by Prions."

111) Richard M. Paddison, M.D. Lecture, Louisiana State University Health Sciences Center, New Orleans, Louisiana,
March 6, 2001,
"Mad Cows, Demented People, and the Biology of Prions."

112) Sterling Drug/Maurice L. Tainter Keynote Lecture, The First Stanford Symposium on Aging: Biology, Disease, and Economics, Stanford, California,
March 13, 2001,
"Age Dependent Neurodegenerative Disease–Lessons from Studies of Prions."

113) Lewis Weinstein Lecture, 39th Annual Meeting of the American Society of Neuroradiology, Boston, Massachusetts,
April 23, 2001,
"Demented People, Mad Cows, and the Biology of Prions."

114) Rodney Porter Memorial Lecture, Oxford University Museum of Natural History, University of Oxford, Oxford, England,
May 31, 2001,
"The Mad Cow Crisis."

115) Lección Conmemorativa
"E. Ortiz de Landázuri",
Clínica Universitaria, Facultad de Medicina, Universidad de Navarra, Pamplona, Spain,
June 4, 2001,
"Mad Cows, Demented People and the Biology of Prions."

116) Tadeusz J. Wiktor Memorial Lecture, The Wistar Institute, University of Pennsylvania, Philadelphia, Pennsylvania,
November 1, 2001,
"Mad Cow, Demented People, and the Biology of Neurodegeneration."

117) Neal Nathanson Lectureship, School of Medicine, University of Pennsylvania, Philadelphia, Pennsylvania,
February 13, 2002,
"Prion Biology and New Approaches to Therapeutics."

118) Thomas Dent Mütter Lecture, The College of Physicians of Philadelphia, Philadelphia, Pennsylvania,
February 14, 2002,
"Prion Biology and New Approaches to Therapeutics."

119) Marian E. Koshland Seminar Series, Department of Molecular and Cell Biology, University of California, Berkeley, California,
May 14, 2002,
"Prion Biology and Therapeutics."

120) Oshman Efron Guest Lecture, Baylor College of Medicine, Houston, Texas,
September 26, 2002,
"Prion Biology, Demented People, and Mad Cows: From Slow Viruses to Misfolded Proteins."

121) Lawson Wilkins Lecture, Lawson Wilkins Pediatric Endocrine Society Annual Meeting, San Francisco, California,
May 1, 2004,
"The Mad Cows That Changed America."

122) Robert Marshak Distinguished Lectureship in Veterinary Medicine, University of Pennsylvania School of Veterinary Medicine Faculty Research Retreat 2004, Bryn Mawr, Pennsylvania,
June 18, 2004,
"The Mad Cows that Changed America."

123) Raymond D. Adams Lecture, 129th Annual Meeting of the American Neurological Association, Toronto, Ontario, Canada,
October 6, 2004,
"Mad Cows and Synthetic Prions."

124) Robert Wartenberg Lecture, American Academy of Neurology 57th Annual Meeting, Miami Beach, Florida,
April 12, 2005,
"Synthetic Prions: Diagnosis and Treatment of Prion Disease"

125) 84th Annual Beaumont Lecture, Wayne County Medical Society, Troy, Michigan,
October 28, 2005,
"Synthetic Prions, Mad Cows, and the Struggle Between Heresy and Orthodoxy in Science."

126) Winford P. Larson Lecture, University of Minnesota Department of Microbiology, Minneapolis, Minnesota,
April 17, 2006,
"Synthetic Prions, Mad Cows, and Scientific Heresy."

127) 3rd Annual Sid Gilman and Carol Barbour Lecture in Experimental Neurology, University of Michigan Health System, Ann Arbor, Michigan,

April 19, 2006,
"Synthetic Prions, Mad Cows, and Scientific Heresy."

128) Sterling B. Hendricks Memorial Lectureship, 232nd American Chemical Society Meeting, San Francisco, California,
September 13, 2006,
"Synthetic Prions, Mad Cows, and Scientific Heresy."

129) Russell and Elizabeth Bundy Visiting Professorship in Honor of Dr. Ross M. Tucker, Mayo Clinic, Jacksonville, Florida,
October 10, 2006,
"Synthetic Prions, Mad Cows, and Scientific Heresy."

130) Dr. Robert B. Church Lecture in Biotechnology, University of Calgary, Calgary, Alberta, Canada,
October 25, 2006,
"Mad Cows, Prion Diagnostics, and Scientific Heresy."

131) Ernst Knobil Distinguished Lecture, University of Texas, Houston, Texas,
November 30, 2006,
"Synthetic Prions, Mad Cows and Scientific Heresy."

132) Twenty-seventh T. S. Srinivasan Endowment Oration, Indian Institute of Technology – Madras, Chennai, India,
February 17, 2007,
"On Viruses, Genes and Mad Cows – Lessons from Prion Disease."

133) Fourth Annual Robert A. Fishman Visiting Professor of Neurology Lecture, University of California, San Francisco, California,
April 23, 2007,
"Prions, Mad Cows and Dementing Diseases."

134) 12th Annual Distinguished Visiting Scientist Lecture, Feinstein Institute for Medical Research, Manhasset,
New York,
June 20, 2007,
"Prions, Mad Cows and Dementing Diseases."

135) Leverhulme Lectures, Imperial College, London, England,
January 17, 2008,
"Discovering Prions – Some Personal Reflections. Looking For a Way Out of the Fog (1972–1978)."
January 24, 2008,
"Discovering Prions – Some Personal Reflections. Searching for a Virus and Finding Only Protein (1978–1987)."
February 28, 2008,
"Discovering Prions – Some Personal Reflections. The Reality of Prions (1988–2007)."

136) Nobel Conversations: discovering the unexpected, Imperial College, London, England,
June 17, 2008,
"Prions – A New Principle of Infection."

137) Elizabeth Rogers Lecture in Geriatric Medicine, Johns Hopkins Medical Institutions, Baltimore, Maryland,
November 10, 2008,
"Neurodegenerative Diseases of Aging."

138) Peter W. Lampert Memorial Lecture, University of California San Diego, Department of Pathology, San Diego, California,
November 21, 2008,
"The Neurobiology of Prion Diseases."

139) Society for General Microbiology Prize Medal Lecture, Harrogate, England,
April 1, 2009,
"Prion Biology and Diseases."

140) Gerald and Sally DeNardo Lectureship Public Lecture, Santa Clara University, College of Arts and Sciences, Santa Clara, California,
April 15, 2009,
"The Scourge of Alzheimer's and Parkinson's and Prion Diseases."

141) Gerald and Sally DeNardo Lectureship Public Lecture, Santa Clara University, College of Arts and Sciences, Santa Clara, California,
April 16, 2009,
"Discovering Prions.

142) National Institutes of Health Distinguished Lecturer in Neuroscience and Aging, Baltimore, Maryland,
May 20, 2009,
"Design and Construction of Prion Strains."

143) Robert E. Gross Lecture, American Pediatric Surgery Association Annual Conference, Fajardo, Puerto Rico,
May 29, 2009,
"Designer Prions and a Quest for Therapy."

144) Presidential Eastman Medal of the University of Rochester, Distinguished Neuroscience Speaker Series, Rochester, New York,
December 8, 2009,
"Design and Construction of Synthetic Prions."

145) 1st Annual Anthony Fink Lecture, University of California Santa Cruz, Santa Cruz, California,
October 15, 2010,
"Increasing Evidence that Prions Cause Most Neurodegenerative Diseases."

146) 1st Annual Charles J. Epstein, M.D. Lecture, Buck Institute for Research on Aging, Novato, California,
February 11, 2011,
"The Spectrum of Prion-Like Diseases and the Quest for Therapeutics."

147) 29th Annual Rita and Taft Schreiber Lectureship in Transfusion Medicine, Cedars Sinai Medical Center, Los Angeles, California,
April 1, 2011,
"Pathogenesis of Prion Disease and Elusive Therapies for Neurodegenerative Diseases."

148) 2nd Annual Penn Chemistry Distinguished Alumni Award Symposium honoring Stanley B. Prusiner, University of Pennsylvania, Philadelphia, Pennsylvania,
May 3, 2011,
"The Biochemistry of Prions"

149) 5th Annual Sackler Lecture, Yale University Program for Cellular Neuroscience, Neurodegeneration and Repair, New Haven, Connecticut,
May 5, 2011,
"The Spectrum of Prion Like Diseases and the Quest for Therapeutics."

150) 3rd Annual Presidential Lecture, Uniformed Services University Research Week, Uniformed Services University, Bethesda, Maryland,
May 18, 2011,
"Therapeutics Approaches to Neurodegeneration in Head Trauma."

151) 2012 Presidential Distinguished Lecture, The University of Texas Health Science Center at San Antonio, San Antonio, Texas,
November 2, 2012,
"Brain Injuries: Soldier, Football Players and Older People."

152) Distinguished Neuroscience Lecture, Nebraska Neuroscience Alliance, Omaha, Nebraska,
December 13, 2012,
"Brain Injuries: Soldier, Football Players and Alzheimer's Victims."

153) Dammin Lecture, Brigham and Women's Hospital, Boston, Massachusetts,
January 24, 2013,
"Unifying Role for Prions in Neurodegeneration."

154) USC Irene McCulloch Distinguished Lecturer in Neuroscience, Keck School of Medicine of USC, Los Angeles, California,
March 14, 2013,
"A Unifying Role for Prions in Neurodegeneration."

155) The Ruth K. Broad Foundation Seminar Series on Neurobiology and Disease, Duke University, Durham, North Carolina,
April 23, 2013,
"A Unifying Role for Prions in Neurodegeneration."

156) DeArmond Lecture, 89th Annual Meeting of the American Association of Neuropathologists, Charleston, South Carolina,
June 21, 2013,
"A Unifying Role for Prions in Neurodegenerative Diseases."

157) 5th Annual Michael Nacht Distinguished Lecture in Politics and Public Policy, University of California Berkeley, Berkeley, California,
February 25, 2014,
"Alzheimer's, Parkinson's and Traumatic Brain Injury—Prions, Amyloids and Proteins."

158) Francis Crick Lecture, Laboratory of Molecular Biology, Cambridge University, England,
April 11, 2014,
"Prions Causing Neurodegeneration: A Unifying Etiology and the Quest for Therapeutics."

159) David Packard Lecture, Uniformed Services University of the Health Sciences, Bethesda, Maryland
April 4, 2016,
"Prions Causing CTE and the Discovery of Anti-Tau Prion Therapeutics."

160) Royalty Pharma Investors Day Lecture, Royalty Pharma, New York City, New York,
September 27, 2016,

"Quest toward effective drugs that prevent or slow Alzheimer's and Parkinson's diseases."

161) Alfred G. Gilman Memorial Symposium, University of Texas, Southwestern Medical Center, Dallas, Texas, October 7, 2016,
"Remembering Al Gilman and Ernest Gallo."

162) Michigan State University, Memorial Lecture for Skip Binder, Grand Rapids, Michigan, October 31, 2016,
"Development of the Prion Concept."

163) Nobel Laureate Revisiting Lecture, Karolinska Institute, Stockholm, Sweden, April 4, 2017,
"Quest for prion therapeutics in neurodegenerative diseases."

164) City of Hope Eugene Roberts Memorial Lecture, Duarte, California, January 22, 2018,
"From GABA to Prions."

165) Don Gilden Memorial Lecture, Vail, Colorado, May 8, 2019,
" Discovering Anti-Prion Therapeutics for Alzheimer's and Parkinson's Diseases."

Invited Lectures (1977 – present): Total of 751

Invited Non-Scientific Presentations (1998 – present): Total of 70

PUBLICATIONS

Journal Articles

1) Black-Schaffer, B., Prusiner, S., and Esparza, H. (1965). Tolerance of the vestibular apparatus of the hypothermic hamster to 840G acceleration. Aerospace Med. *36*, 123–126.

2) Prusiner, S.B., Moskovitz, P.A., and Wolfson, S.K., Jr. (1965). Relationship of acidemia to cerebral edema. Arch. Surg. *91*, 902–905.

3) Prusiner, S., and Wolfson, S.K., Jr. (1968). Hypothermic protection against cerebral edema of ischemia. Arch. Neurol. *19*, 623–627.

4) Prusiner, S., Williamson, J.R., Chance, B., and Paddle, B.M. (1968). Pyridine nucleotide changes during thermogenesis in brown fat tissue in vivo. Arch. Biochem. Biophys. *123*, 368–377.

5) Prusiner, S.B., Eisenhardt, R.H., Rylander, E., and Lindberg, O. (1968). The regulation of oxidative metabolism of isolated brown fat cells. Biochem. Biophys. Res. Commun. *30*, 508–515.

6) Lindberg, O., Prusiner, S.B., Cannon, B., Ching, T.M., and Eisenhardt, R.H. (1970). Metabolic control in isolated brown fat cells. Lipids *5*, 204–209.

7) Prusiner, S., and Milner, L. (1970). A rapid radioactive assay for glutamine synthetase, glutaminase, asparagine synthetase, and asparaginase. Anal. Biochem. *37*, 429–438.

8) Prusiner, S. (1970). Spectroscopic evidence for the control of respiration prior to phosphorylation in hamster brown fat cells. J. Biol. Chem. *245*, 382–389.

9) Williamson, J.R., Prusiner, S., Olson, M.S., and Fukami, M. (1970). Control of metabolism in brown adipose tissue. Lipids *5*, 1–14.

10) Prusiner, S.B., Milner, L.S., Long, C.W., and Myers, M.L. (1971). Vacuum manifold for rapid assay of enzymes using radioactive tracers and ion exchange chromatography. Rev. Sci. Instrum. *42*, 493–494.

11) Prusiner, S., Miller, R.E., and Valentine, R. (1972). Adenosine 3':5'-cyclic monophosphate control of the enzymes of glutamine metabolism in *Escherichia coli*. Proc. Natl. Acad. Sci. U.S.A. *69*, 2922–2926.

12) Deuel, T., and Prusiner, S. (1974). Regulation of glutamine synthetase from *Bacillus subtilis* by divalent cations, feedback inhibitors, and L-glutamine. J. Biol. Chem. *249*, 257–264.

13) Prusiner, S. (1975). Regulation of glutaminase levels in Escherichia coli. J. Bacteriol. *123*, 992–999.

14) Prusiner, S., Davis, J.N., and Stadtman, E.R. (1976). Regulation of glutaminase B in *Escherichia coli*. I. Purification, properties, and cold lability. J. Biol. Chem. *251*, 3447–3456.

15) Prusiner, S., Doak, C.W., and Kirk, G. (1976). A novel mechanism for group translocation: substrate-product reutilization by gamma-glutamyl transpeptidase in peptide and amino acid transport. J. Cell Physiol. *89*, 853–863.

16) Prusiner, S., and Stadtman, E.R. (1976a). Regulation of glutaminase B in *Escherichia coli*. II. Modulation of activity by carboxylate and borate ions. J. Biol. Chem. *251*, 3457–3462.

17) Prusiner, S.B., and Stadtman, E.R. (1976b). Regulation of glutaminase B in *Escherichia coli*. III. Control by nucleotides and divalent cations. J. Biol. Chem. *251*, 3463–3469.

18) Kirk, G., and Prusiner, S.B. (1977). Comparative studies on membranes from bovine choroid plexus and rat kidney cortex. Life Sci. *21*, 833–840.

19) Prusiner, S.B., Hadlow, W.J., Eklund, C.M., and Race, R.E. (1977). Sedimentation properties of the scrapie agent. Proc. Natl. Acad. Sci. U.S.A. *74*, 4656–4660.

20) Raskin, N.H., and Prusiner, S. (1977). Carotidynia. Neurology *27*, 43–46.

21) Baringer, J.R., and Prusiner, S.B. (1978). Experimental scrapie in mice: ultrastructural observations. Ann. Neurol. *4*, 205–211.

22) Garfin, D.E., Stites, D.P., Perlman, J.D., Cochran, S.P., and Prusiner, S.B. (1978a). Mitogen stimulation of splenocytes from mice infected with scrapie agent. J. Infect. Dis. *138*, 396–400.

23) Garfin, D.E., Stites, D.P., Zitnik, L.A., and Prusiner, S.B. (1978b). Suppression of polyclonal B cell activation in scrapie-infected C3H/HeJ mice. J. Immunol. *120*, 1986–1990.

24) Hittelman, K., Mamelok, R., and Prusiner, S.B. (1978). Preservation by freezing of glucose and alanine transport into kidney membrane vesicles. Anal. Biochem. *89*, 324–331.

25) Prusiner, P.E., and Prusiner, S.B. (1978a). Partial purification and kinetics of gamma-glutamyl transpeptidase from bovine choroid plexus. J. Neurochem. *30*, 1253–1259.

26) Prusiner, P.E., and Prusiner, S.B. (1978b). Modulation of gamma-glutamyl transpeptidase activity from bovine choroid plexus. J. Neurochem. *30*, 1261–1267.

27) Prusiner, S.B. (1978). An approach to the isolation of biological particles using sedimentation analysis. J. Biol. Chem. *253*, 916–921.

28) Prusiner, S.B., Garfin, D.E., Cochran, S.P., Baringer, J.R., Hadlow, W.J., Eklund, C.M., and Race, R.E. (1978a). Evidence for hydrophobic domains on the surface of the scrapie agent. Trans. Am. Neurol. Assoc. *103*, 62–64.

29) Prusiner, S.B., Hadlow, W.J., Eklund, C.M., Race, R.E., and Cochran, S.P. (1978b). Sedimentation characteristics of the scrapie agent from murine spleen and brain. Biochemistry *17*, 4987–4992.

30) Prusiner, S.B., Hadlow, W.J., Garfin, D.E., Cochran, S.P., Baringer, J.R., Race, R.E., and Eklund, C.M. (1978c). Partial purification and evidence for multiple molecular forms of the scrapie agent. Biochemistry *17*, 4993–4997.

31) Chatigny, M.A., Ishimaru, K., Dunn, S., Eagleson, J.A., and Prusiner, S.B. (1979). Evaluation of a class III biological safety cabinet for enclosure of an ultracentrifuge. Appl. Environ. Microbiol. *38*, 934–939.

32) Chatigny, M.A., and Prusiner, S.B. (1980). Biohazards of investigations on the transmissible spongiform encephalopathies. Rev. Infect. Dis. *2*, 713–724.

33) Hadlow, W.J., Prusiner, S.B., Kennedy, R.C., and Race, R.E. (1980). Brain tissue from persons dying of Creutzfeldt-Jakob disease causes scrapie-like encephalopathy in goats. Ann. Neurol. *8*, 628–631.

34) Mamelok, R., Groth, D., and Prusiner, S.B. (1980). Separation of membrane-bound g-glutamyl transpeptidase from brush border transport and enzyme activities. Biochemistry *19*, 2367–2373.

35) Prusiner, S.B., Garfin, D.E., Cochran, S.P., McKinley, M.P., Groth, D.F., Hadlow, W.J., Race, R.E., and Eklund, C.M. (1980a). Experimental scrapie in the mouse: electrophoretic and sedimentation properties of the partially purified agent. J. Neurochem. *35*, 574–582.

36) Prusiner, S.B., Groth, D.F., Bildstein, C., Masiarz, F.R., McKinley, M.P., and Cochran, S.P. (1980b). Electrophoretic properties of the scrapie agent in agarose gels. Proc. Natl. Acad. Sci. U.S.A. *77*, 2984–2988.

37) Prusiner, S.B., Groth, D.F., Cochran, S.P., Masiarz, F.R., McKinley, M.P., and Martinez, H.M. (1980c). Molecular properties, partial purification, and assay by incubation period measurements of the hamster scrapie agent. Biochemistry *21*, 4883–4891.

38) Prusiner, S.B., Groth, D.F., Cochran, S.P., McKinley, M.P., and Masiarz, F.R. (1980d). Gel electrophoresis and glass permeation chromatography of the hamster scrapie agent after enzymatic digestion and detergent extraction. Biochemistry *19*, 4892–4898.

39) Baringer, J.R., Prusiner, S.B., and Wong, J. (1981). Scrapie-associated particles in post-synaptic processes. Further ultrastructural studies. J. Neuropathol. Exp. Neurol. *40*, 281–288.

40) Crook, R.B., Kasagami, H., and Prusiner, S.B. (1981). Culture and characterization of epithelial cells from bovine choroid plexus. J. Neurochem. *37*, 845–854.

41) Hogan, R.N., Baringer, J.R., and Prusiner, S.B. (1981). Progressive retinal degeneration in scrapie-infected hamsters: a light and electron microscopic analysis. Lab. Invest. *44*, 34–42.

42) Mamelok, R.D., Macrae, D.R., Benet, L.Z., and Prusiner, S.B. (1981a). Membrane populations of bovine choroid plexus: separation by density gradient centrifugation in modified colloidal silica. J. Neurochem. *37*, 768–774.

43) Mamelok, R.D., Macrae, D.R., Hittelman, K., Hoefer, P.J., and Prusiner, S.B. (1981b). Kinetics of D-glucose transport into renal membrane vesicles - measurements using a vacuum manifold apparatus. J. Biochem. Biophys. Methods *4*, 147–153.

44) McKinley, M.P., Masiarz, F.R., and Prusiner, S.B. (1981). Chemical modification of a scrapie agent protein by diethylpyrocarbonate. Trans. Am. Neurol. Assoc. *106*, 293–296.

45) McKinley, M.P., Masiarz, F.R., and Prusiner, S.B. (1981). Reversible chemical modification of the scrapie agent. Science *214*, 1259–1261.

46) Prusiner, S.B. (1981). Disorders of glutamate metabolism and neurological dysfunction. Annu. Rev. Med. *32*, 521–542.

47) Prusiner, S.B., Groth, D.F., McKinley, M.P., Cochran, S.P., Bowman, K.A., and Kasper, K.C. (1981a). Thiocyanate and hydroxyl ions inactivate the scrapie agent. Proc. Natl. Acad. Sci. U.S.A. *78*, 4606–4610.

48) Prusiner, S.B., McKinley, M.P., Groth, D.F., Bowman, K.A., Mock, N.I., Cochran, S.P., and Masiarz, F.R. (1981b). Scrapie agent contains a hydrophobic protein. Proc. Natl. Acad. Sci. U.S.A. *78*, 6675–6679.

49) Bolton, D.C., McKinley, M.P., and Prusiner, S.B. (1982). Identification of a protein that purifies with the scrapie prion. Science *218*, 1309–1311.

50) Diener, T.O., McKinley, M.P., and Prusiner, S.B. (1982). Viroids and prions. Proc. Natl. Acad. Sci. U.S.A. *79*, 5220–5224.

51) Kasper, K.C., Stites, D.P., Bowman, K.A., Panitch, H., and Prusiner, S.B. (1982). Immunological studies of scrapie infection. J. Neuroimmunol. *3*, 187–201.

52) Prusiner, S.B. (1982a). Novel proteinaceous infectious particles cause scrapie. Science *216*, 136–144.

53) Prusiner, S.B. (1982b). Research on scrapie. Lancet *320*, 494–495.

54) Prusiner, S.B. (1982c). On prions causing dementia - molecular studies of the scrapie agent. Curr. Neurol. *4*, 201–224.

55) Prusiner, S.B., Bolton, D.C., Groth, D.F., Bowman, K.A., Cochran, S.P., and McKinley, M.P. (1982a). Further purification and characterization of scrapie prions. Biochemistry *21*, 6942–6950.

56) Prusiner, S.B., Cochran, S.P., Groth, D.F., Downey, D.E., Bowman, K.A., and Martinez, H.M. (1982b). Measurement of the scrapie agent using an incubation time interval assay. Ann. Neurol. *11*, 353–358.

57) Prusiner, S.B., Gajdusek, D.C., and Alpers, M.P. (1982c). Kuru with incubation periods exceeding two decades. Ann. Neurol. *12*, 1–9.

58) Baringer, J.R., Bowman, K.A., and Prusiner, S.B. (1983). Replication of the scrapie agent in hamster brain precedes neuronal vacuolation. J. Neuropathol. Exp. Neurol. *42*, 539–547.

59) Hogan, R.N., Kingsbury, D.T., Baringer, J.R., and Prusiner, S.B. (1983). Retinal degeneration in experimental Creutzfeldt-Jakob disease. Lab. Invest. *49*, 708–715.

60) Kingsbury, D.T., Kasper, K.C., Stites, D.P., Watson, J.D., Hogan, R.N., and Prusiner, S.B. (1983). Genetic control of scrapie and Creutzfeldt-Jakob disease in mice. J. Immunol. *131*, 491–496.

61) McKinley, M.P., Bolton, D.C., and Prusiner, S.B. (1983a). A protease-resistant protein is a structural component of the scrapie prion. Cell *35*, 57–62.

62) McKinley, M.P., Masiarz, F.R., Isaacs, S.T., Hearst, J.E., and Prusiner, S.B. (1983b). Resistance of the scrapie agent to inactivation by psoralens. Photochem. Photobiol. *37*, 539–545.

63) Prusiner, S.B., McKinley, M.P., Bowman, K.A., Bolton, D.C., Bendheim, P.E., Groth, D.F., and Glenner, G.G. (1983). Scrapie prions aggregate to form amyloid-like birefringent rods. Cell *35*, 349–358.

64) Roach, A., Boylan, K., Horvath, S., Prusiner, S.B., and Hood, L.E. (1983). Characterization of cloned cDNA rat myelin basic protein: absence of expression in brain of shiverer mutant mice. Cell *34*, 799–806.

65) Bendheim, P.E., Barry, R.A., DeArmond, S.J., Stites, D.P., and Prusiner, S.B. (1984). Antibodies to a scrapie prion protein. Nature *310*, 418–421.

66) Bolton, D.C., McKinley, M.P., and Prusiner, S.B. (1984). Molecular characteristics of the major scrapie prion protein. Biochemistry *23*, 5898–5906.

67) Crook, R.B., Farber, M.B., and Prusiner, S.B. (1984). Hormones and neurotransmitters control cyclic AMP metabolism in choroid plexus epithelial cells. J. Neurochem. *42*, 340–350.

68) Friedland, R.P., Prusiner, S.B., Jagust, W.J., Budinger, T.F., and Davis, R.L. (1984). Bitemporal hypometabolism in Creutzfeldt-Jakob disease measured by positron emission tomography with [^{18}F]-2-fluorodeoxyglucose. J. Comput. Assist. Tomogr. *8*, 978–981.

69) Prusiner, S.B. (1984a). Prions. Sci. Am. *251*, 50–59.

70) Prusiner, S.B. (1984b). Prions: novel infectious pathogens. Adv. Virus Res. *29*, 1–56.

71) Prusiner, S.B. (1984c). Some speculations about prions, amyloid, and Alzheimer's disease. N. Engl. J. Med. *310*, 661–663.

72) Prusiner, S.B., Groth, D.F., Bolton, D.C., Kent, S.B., and Hood, L.E. (1984). Purification and structural studies of a major scrapie prion protein. Cell *38*, 127–134.

73) Barry, R.A., McKinley, M.P., Bendheim, P.E., Lewis, G.K., DeArmond, S.J., and Prusiner, S.B. (1985). Antibodies to the scrapie protein decorate prion rods. J. Immunol. *135*, 603–613.

74) Bendheim, P.E., Bockman, J.M., McKinley, M.P., Kingsbury, D.T., and Prusiner, S.B. (1985). Scrapie and Creutzfeldt-Jakob disease prion proteins share physical properties and antigenic determinants. Proc. Natl. Acad. Sci. U.S.A. *82*, 997–1001.

75) Bockman, J.M., Kingsbury, D.T., McKinley, M.P., Bendheim, P.E., and Prusiner, S.B. (1985). Creutzfeldt-Jakob disease prion proteins in human brains. N. Engl. J. Med. *312*, 73–78.

76) Bolton, D.C., Meyer, R.K., and Prusiner, S.B. (1985). Scrapie PrP 27–30 is a sialoglycoprotein. J. Virol. *53*, 596–606.

77) DeArmond, S.J., McKinley, M.P., Barry, R.A., Braunfeld, M.B., McColloch, J.R., and Prusiner, S.B. (1985). Identification of prion amyloid filaments in scrapie-infected brain. Cell *41*, 221–235.

78) Monteiro, M.L.R., Swanson, R.A., Coppeto, J.R., Cuneo, R.A., DeArmond, S.J., and Prusiner, S.B. (1985). A microangiopathic syndrome of encephalopathy, hearing loss, and retinal arteriolar occlusions. Neurology *35*, 1113–1121.

79) Oesch, B., Westaway, D., Wälchli, M., McKinley, M. P., Kent, S. B. H., Aebersold, R., Barry, R. A., Tempst, P., Teplow, D. B., Hood, L. E., Prusiner, S. B., and Weissmann, C. (1985). A cellular gene encodes scrapie PrP 27–30 protein. Cell *40*, 735–746.

80) Prusiner, S.B., Barry, R.A., McKinley, M.P., Bellinger, C.G., Meyer, R.K., and DeArmond, S.J. (1985a). Scrapie and Creutzfeldt-Jakob disease prions. Microbiol. Sci. *2*, 33–39.

81) Prusiner, S.B., Cochran, S.P., and Alpers, M.P. (1985b). Transmission of scrapie in hamsters. J. Infect. Dis. *152*, 971–978.

82) Prusiner, S.B., and Kingsbury, D.T. (1985). Prions - infectious pathogens causing the spongiform encephalopathies. CRC Crit. Rev. Clin. Neurobiol. *1*, 181–200.

83) Takahashi, N., Roach, A., Teplow, D.B., Prusiner, S.B., and Hood, L. (1985). Cloning and characterization of the myelin basic protein gene from mouse: one gene can encode both 14 kd and 18.5 kd MBPs by alternate use of exons. Cell *42*, 139–148.

84) Barry, R.A., Kent, S.B.H., McKinley, M.P., Meyer, R.K., DeArmond, S.J., Hood, L.E., and Prusiner, S.B. (1986). Scrapie and cellular prion proteins share polypeptide epitopes. J. Infect. Dis. *153*, 848–854.

85) Barry, R.A., and Prusiner, S.B. (1986). Monoclonal antibodies to the cellular and scrapie prion proteins. J. Infect. Dis. *154*, 518–521.

86) Basler, K., Oesch, B., Scott, M., Westaway, D., Wälchli, M., Groth, D.F., McKinley, M.P., Prusiner, S.B., and Weissmann, C. (1986). Scrapie and cellular PrP isoforms are encoded by the same chromosomal gene. Cell *46*, 417–428.

87) Carlson, G.A., Kingsbury, D.T., Goodman, P.A., Coleman, S., Marshall, S.T., DeArmond, S., Westaway, D., and Prusiner, S.B. (1986). Linkage of prion protein and scrapie incubation time genes. Cell *46*, 503–511.

88) Crook, R.B., Farber, M.B., and Prusiner, S.B. (1986). H_2 histamine receptors on the epithelial cells of choroid plexus. J. Neurochem. *46*, 489–493.

89) Crook, R.B., and Prusiner, S.B. (1986). Vasoactive intestinal peptide stimulates cyclic AMP in choroid plexus epithelial cells. Brain Res. *384*, 138–144.

90) Hogan, R.N., Bowman, K.A., Baringer, J.R., and Prusiner, S.B. (1986). Replication of scrapie prions in hamster eyes precedes retinal degeneration. Ophthalmic Res. *18*, 230–235.

91) Kitamoto, T., Tateishi, J., Tashima, I., Takeshita, I., Barry, R.A., DeArmond, S.J., and Prusiner, S.B. (1986). Amyloid plaques in Creutzfeldt-Jakob disease stain with prion protein antibodies. Ann. Neurol. *20*, 204–208.

92) Kretzschmar, H.A., Prusiner, S.B., Stowring, L.E., and DeArmond, S.J. (1986a). Scrapie prion proteins are synthesized in neurons. Am. J. Pathol. *122*, 1–5.

93) Kretzschmar, H.A., Stowring, L.E., Westaway, D., Stubblebine, W.H., Prusiner, S.B., and DeArmond, S.J. (1986b). Molecular cloning of a human prion protein cDNA. DNA *5*, 315–324.

94) McKinley, M.P., Braunfeld, M.B., Bellinger, C.G., and Prusiner, S.B. (1986). Molecular characteristics of prion rods purified from scrapie-infected hamster brains. J. Infect. Dis. *154*, 110–120.

95) Meyer, R.K., McKinley, M.P., Bowman, K.A., Braunfeld, M.B., Barry, R.A., and Prusiner, S.B. (1986). Separation and properties of cellular and scrapie prion proteins. Proc. Natl. Acad. Sci. U.S.A. *83*, 2310–2314.

96) Prusiner, S.B. (1986). Prions are novel infectious pathogens causing scrapie and Creutzfeldt-Jakob disease. Bioessays *5*, 281–286.

97) Roberts, G.W., Lofthouse, R., Brown, R., Crow, T.J., Barry, R.A., and Prusiner, S.B. (1986). Prion-protein immunoreactivity in human transmissible dementias. N. Engl. J. Med. *315*, 1231–1233.

98) Sparkes, R. S., Simon, M., Cohn, V. H., Fournier, R. E. K., Lem, J., Klisak, I., Heinzmann, C., Blatt, C., Lucero, M., Mohandas, T., DeArmond, S. J., Westaway, D., Prusiner, S. B., and Weiner, L. P. (1986). Assignment of the human and mouse prion protein genes to homologous chromosomes. Proc. Natl. Acad. Sci. U.S.A. *83*, 7358–7362.

99) Westaway, D., and Prusiner, S.B. (1986). Conservation of the cellular gene encoding the scrapie prion protein. Nucleic Acids Res. *14*, 2035–2044.

100) Bazan, J.F., Fletterick, R.J., McKinley, M.P., and Prusiner, S.B. (1987a). Predicted secondary structure and membrane topology of the scrapie prion protein. Protein Eng. *1*, 125–135.

101) Bazan, J.F., Fletterick, R.J., and Prusiner, S.B. (1987b). Lack of similarity between AIDS virus and scrapie protein genes. Nature *325*, 581.

102) Bellinger-Kawahara, C., Cleaver, J.E., Diener, T.O., and Prusiner, S.B. (1987a). Purified scrapie prions resist inactivation by UV irradiation. J. Virol. *61*, 159–166.

103) Bellinger-Kawahara, C., Diener, T.O., McKinley, M.P., Groth, D.F., Smith, D.R., and Prusiner, S.B. (1987b). Purified scrapie prions resist inactivation by procedures that hydrolyze, modify, or shear nucleic acids. Virology *160*, 271–274.

104) Bockman, J.M., Prusiner, S.B., Tateishi, J., and Kingsbury, D.T. (1987). Immunoblotting of Creutzfeldt-Jakob disease prion proteins: host species-specific epitopes. Ann. Neurol. *21*, 589–595.

105) Boylan, K.B., Takahashi, N., Diamond, M., Hood, L.E., and Prusiner, S.B. (1987). DNA length polymorphism located 5' to the human myelin basic protein gene. Am. J. Hum. Genet. *40*, 387–400.

106) Braun, M.J., Gonda, M.A., George, D.G., Bazan, J.F., Fletterick, R.J., and Prusiner, S.B. (1987). The burden of proof in linking AIDS to scrapie. Nature *330*, 525–526.

107) DeArmond, S.J., Mobley, W.C., DeMott, D.L., Barry, R.A., Beckstead, J.H., and Prusiner, S.B. (1987). Changes in the localization of brain prion proteins during scrapie infection. Neurology *37*, 1271–1280.

108) Gabizon, R., McKinley, M.P., and Prusiner, S.B. (1987). Purified prion proteins and scrapie infectivity copartition into liposomes. Proc. Natl. Acad. Sci. U.S.A. *84*, 4017–4021.

109) Hay, B., Barry, R.A., Lieberburg, I., Prusiner, S.B., and Lingappa, V.R. (1987a). Biogenesis and transmembrane orientation of the cellular isoform of the scrapie prion protein. Mol. Cell. Biol. *7*, 914–920.

110) Hay, B., Prusiner, S.B., and Lingappa, V.R. (1987b). Evidence for a secretory form of the cellular prion protein. Biochemistry *26*, 8110–8115.

111) Hogan, R.N., Baringer, J.R., and Prusiner, S.B. (1987). Scrapie infection diminishes spines and increases varicosities of dendrites in hamsters: a quantitative Golgi analysis. J. Neuropathol. Exp. Neurol. *46*, 461–473.

112) Kitamoto, T., Ogomori, K., Tateishi, J., and Prusiner, S.B. (1987). Formic acid pretreatment enhances immunostaining of cerebral and systemic amyloids. Lab. Invest. *57*, 230–236.

113) McKinley, M.P., Hay, B., Lingappa, V.R., Lieberburg, I., and Prusiner, S.B. (1987). Developmental expression of prion protein gene in brain. Dev. Biol. *121*, 105–110.

114) Prusiner, S.B. (1987a). Prions causing degenerative neurological diseases. Annu. Rev. Med. *38*, 381–398.

115) Prusiner, S.B. (1987b). Prion diseases and central nervous system degeneration. Clin. Res. *35*, 177–191.

116) Prusiner, S.B. (1987c). Prions and neurodegenerative diseases. N. Engl. J. Med. *317*, 1571–1581.

117) Prusiner, S.B., and DeArmond, S.J. (1987). Biology of disease: Prions causing nervous system degeneration. Lab. Invest. *56*, 349–363.

118) Prusiner, S.B., Gabizon, R., and McKinley, M.P. (1987). On the biology of prions. Acta Neuropathol. (Berl.) *72*, 299–314.

119) Stahl, N., Borchelt, D.R., Hsiao, K., and Prusiner, S.B. (1987). Scrapie prion protein contains a phosphatidylinositol glycolipid. Cell *51*, 229–240.

120) Westaway, D., Goodman, P.A., Mirenda, C.A., McKinley, M.P., Carlson, G.A., and Prusiner, S.B. (1987). Distinct prion proteins in short and long scrapie incubation period mice. Cell *51*, 651–662.

121) Wiley, C.A., Burrola, P.G., Buchmeier, M.J., Wooddell, M.K., Barry, R.A., Prusiner, S.B., and Lampert, P.W. (1987). Immuno-gold localization of prion filaments in scrapie-infected hamster brains. Lab. Invest. *57*, 646–656.

122) Barry, R.A., Vincent, M.T., Kent, S.B.H., Hood, L.E., and Prusiner, S.B. (1988). Characterization of prion proteins with monospecific antisera to synthetic peptides. J. Immunol. *140*, 1188–1193.

123) Bellinger-Kawahara, C.G., Kempner, E., Groth, D.F., Gabizon, R., and Prusiner, S.B. (1988). Scrapie prion liposomes and rods exhibit target sizes of 55,000 Da. Virology *164*, 537–541.

124) Butler, D.A., Scott, M.R.D., Bockman, J.M., Borchelt, D.R., Taraboulos, A., Hsiao, K.K., Kingsbury, D.T., and Prusiner, S.B. (1988). Scrapie-infected murine neuroblastoma cells produce protease-resistant prion proteins. J. Virol. *62*, 1558–1564.

125) Carlson, G.A., Goodman, P.A., Lovett, M., Taylor, B.A., Marshall, S.T., Peterson-Torchia, M., Westaway, D., and Prusiner, S.B. (1988). Genetics and polymorphism of the mouse prion gene complex: control of scrapie incubation time. Mol. Cell. Biol. *8*, 5528–5540.

126) Gabizon, R., McKinley, M.P., Groth, D., and Prusiner, S.B. (1988a). Immunoaffinity purification and neutralization of scrapie prion infectivity. Proc. Natl. Acad. Sci. U.S.A. *85*, 6617–6621.

127) Gabizon, R., McKinley, M.P., Groth, D.F., Kenaga, L., and Prusiner, S.B. (1988b). Properties of scrapie prion protein liposomes. J. Biol. Chem. *263*, 4950–4955.

128) Hsiao, K.K., Westaway, D.A., and Prusiner, S.B. (1988). An amino acid substitution in the prion protein of ataxic Gerstmann-Sträussler syndrome. Am. J. Hum. Genet. *43*, A87.

129) Mobley, W.C., Neve, R.L., Prusiner, S.B., and McKinley, M.P. (1988). Nerve growth factor increases mRNA levels for the prion protein and the b-amyloid protein precursor in developing hamster brain. Proc. Natl. Acad. Sci. U.S.A. *85*, 9811–9815.

130) Prusiner, S.B. (1988). Molecular structure, biology and genetics of prions. Adv. Virus Res. *35*, 83–136.

131) Roberts, G.W., Lofthouse, R., Allsop, D., Landon, M., Kidd, M., Prusiner, S.B., and Crow, T.J. (1988). CNS amyloid proteins in neurodegenerative diseases. Neurology *38*, 1534–1540.

132) Scott, M.R.D., Butler, D.A., Bredesen, D.E., Wälchli, M., Hsiao, K.K., and Prusiner, S.B. (1988). Prion protein gene expression in cultured cells. Protein Eng. *2*, 69–76.

133) Turk, E., Teplow, D.B., Hood, L.E., and Prusiner, S.B. (1988). Purification and properties of the cellular and scrapie hamster prion proteins. Eur. J. Biochem. *176*, 21–30.

134) Carlson, G.A., Westaway, D., DeArmond, S.J., Peterson-Torchia, M., and Prusiner, S.B. (1989). Primary structure of prion protein may modify scrapie isolate properties. Proc. Natl. Acad. Sci. U.S.A. *86*, 7475–7479.

135) Endo, T., Groth, D., Prusiner, S.B., and Kobata, A. (1989). Diversity of oligosaccharide structures linked to asparagines of the scrapie prion protein. Biochemistry *28*, 8380–8388.

136) Haraguchi, T., Fisher, S., Olofsson, S., Endo, T., Groth, D., Tarantino, A., Borchelt, D. R., Teplow, D., Hood, L., Burlingame, A., Lycke, E., Kobata, A., and Prusiner, S. B. (1989). Asparagine-linked glycosylation of the scrapie and cellular prion proteins. Arch. Biochem. Biophys. *274*, 1–13.

137) Hsiao, K., Baker, H.F., Crow, T.J., Poulter, M., Owen, F., Terwilliger, J.D., Westaway, D., Ott, J., and Prusiner, S.B. (1989a). Linkage of a prion protein missense variant to Gerstmann-Sträussler syndrome. Nature *338*, 342–345.

138) Hsiao, K.K., Doh-ura, K., Kitamoto, T., Tateishi, J., and Prusiner, S.B. (1989b). A prion protein amino acid substitution in ataxic Gerstmann-Sträussler syndrome. Ann. Neurol. *26*, 137.

139) McKinley, M.P., DeArmond, S.J., Torchia, M., Mobley, W.C., and Prusiner, S.B. (1989). Acceleration of scrapie in neonatal Syrian hamsters. Neurology *39*, 1319–1324.

140) Owen, F., Poulter, M., Lofthouse, R., Collinge, J., Crow, T.J., Risby, D., Baker, H.F., Ridley, R.M., Hsiao, K., and Prusiner, S.B. (1989). Insertion in prion protein gene in familial Creutzfeldt-Jakob disease. Lancet *333*, 51–52.

141) Prusiner, S.B. (1989a). Creutzfeldt-Jakob disease and scrapie prions. Alzheimer Dis. Assoc. Disord. *3*, 52–78.

142) Prusiner, S.B. (1989b). Scrapie prions. Annu. Rev. Microbiol. *43*, 345–374.

143) Scott, M., Foster, D., Mirenda, C., Serban, D., Coufal, F., Wälchli, M., Torchia, M., Groth, D., Carlson, G., DeArmond, S. J., Westaway, D., and Prusiner, S. B. (1989). Transgenic mice expressing hamster prion protein produce species-specific scrapie infectivity and amyloid plaques. Cell *59*, 847–857.

144) Snow, A.D., Kisilevsky, R., Willmer, J., Prusiner, S.B., and DeArmond, S.J. (1989). Sulfated glycosaminoglycans in amyloid plaques of prion diseases. Acta Neuropathol. (Berl.) *77*, 337–342.

145) Westaway, D., Carlson, G.A., and Prusiner, S.B. (1989). Unraveling prion diseases through molecular genetics. Trends Neurosci. *12*, 221–227.

146) Baldwin, M.A., Falick, A.M., Gibson, B.W., Prusiner, S.B., Stahl, N., and Burlingame, A.L. (1990a). Tandem mass spectrometry of peptides with N-terminal glutamine: studies on a prion protein peptide. J. Am. Soc. Mass Spectrom. *1*, 258–264.

147) Baldwin, M.A., Stahl, N., Burlingame, A.L., and Prusiner, S.B. (1990b). Structure determination of glycoinsotiol phospholipid anchors by permethylation and tandem mass spectrometry. Methods: A Companion to Methods in Enzymology *1*, 306–314.

148) Baldwin, M.A., Stahl, N., Reinders, L.G., Gibson, B.W., Prusiner, S.B., and Burlingame, A.L. (1990c). Permethylation and tandem mass spectrometry of oligosaccharides having free hexosamine: analysis of the glycoinositol phospholipd anchor glycan from the scrapie prion protein. Anal. Biochem. *191*, 174–182.

149) Borchelt, D.R., Scott, M., Taraboulos, A., Stahl, N., and Prusiner, S.B. (1990). Scrapie and cellular prion proteins differ in their kinetics of synthesis and topology in cultured cells. J. Cell Biol. *110*, 743–752.

150) Boylan, K.B., Takahashi, N., Paty, D.W., Sadovnick, A.D., Diamond, M., Hood, L.E., and Prusiner, S.B. (1990). DNA length polymorphism 5' to the myelin basic protein gene is associated with multiple sclerosis. Ann Neurol *27*, 291–297.

151) Gabizon, R., and Prusiner, S.B. (1990). Prion liposomes. Biochem. J. *266*, 1–14.

152) Hsiao, K., Cass, C., Conneally, P.M., Dloughy, S.R., Hodes, M.E., Farlow, M.R., Ghetti, B., and Prusiner, S.B. (1990a). Atypical Gerstmann-Sträussler-Scheinker syndrome with neurofibrillary tangles: no mutation in the prion protein open-reading-frame in a patient of the Indiana kindred. Neurobiol. Aging *11*, 302.

153) Hsiao, K., Cass, C., Schellenberg, G., Dwine-Gage, E., Bird, T., and Prusiner, S.B. (1990b). Correlation of specific prion protein mutations with different forms of prion diseases. Neurology *40*, 388.

154) Hsiao, K., and Prusiner, S.B. (1990). Inherited human prion diseases. Neurology *40*, 1820–1827.

155) Hsiao, K.K., Scott, M., Foster, D., Groth, D.F., DeArmond, S.J., and Prusiner, S.B. (1990c). Spontaneous neurodegeneration in transgenic mice with mutant prion protein. Science *250*, 1587–1590.

156) Lopez, C.D., Yost, C.S., Prusiner, S.B., Myers, R.M., and Lingappa, V.R. (1990). Unusual topogenic sequence directs prion protein biogenesis. Science *248*, 226–229.

157) Lowenstein, D.H., Butler, D.A., Westaway, D., McKinley, M.P., DeArmond, S.J., and Prusiner, S.B. (1990). Three hamster species with different scrapie incubation times and neuropathological features encode distinct prion proteins. Mol. Cell. Biol. *10*, 1153–1163.

158) Oesch, B., Teplow, D.B., Stahl, N., Serban, D., Hood, L.E., and Prusiner, S.B. (1990). Identification of cellular proteins binding to the scrapie prion protein. Biochemistry *29*, 5848–5855.

159) Prusiner, S.B. (1990). Novel structure and genetics of prions causing neurodegeneration in humans and animals. Biologicals *18*, 247–262.

160) Prusiner, S. B., Scott, M., Foster, D., Pan, K.-M., Groth, D., Mirenda, C., Torchia, M., Yang, S.-L., Serban, D., Carlson, G. A., Hoppe, P. C., Westaway, D., and DeArmond, S. J. (1990). Transgenetic studies implicate interactions between homologous PrP isoforms in scrapie prion replication. Cell *63*, 673–686.

161) Rogers, M., Taraboulos, A., Scott, M., Groth, D., and Prusiner, S.B. (1990). Intracellular accumulation of the cellular prion protein after mutagenesis of its Asn-linked glycosylation sites. Glycobiology *1*, 101–109.

162) Serban, D., Taraboulos, A., DeArmond, S.J., and Prusiner, S.B. (1990). Rapid detection of Creutzfeldt-Jakob disease and scrapie prion proteins. Neurology *40*, 110–117.

163) Snow, A.D., Wight, T.N., Nochlin, D., Koike, Y., Kimata, K., DeArmond, S.J., and Prusiner, S.B. (1990). Immunolocalization of heparan sulfate proteoglycans to the prion protein amyloid plaques of Gerstmann-Sträussler syndrome, Creutzfeldt-Jakob disease and scrapie. Lab. Invest. *63*, 601–611.

164) Stahl, N., Baldwin, M.A., Burlingame, A.L., and Prusiner, S.B. (1990a). Identification of glycoinositol phospholipid linked and truncated forms of the scrapie prion protein. Biochemistry *29*, 8879–8884.

165) Stahl, N., Borchelt, D.R., and Prusiner, S.B. (1990b). Differential release of cellular and scrapie prion proteins from cellular membranes by phosphatidylinositol-specific phospholipase C. Biochemistry *29*, 5405–5412.

166) Taraboulos, A., Rogers, M., Borchelt, D.R., McKinley, M.P., Scott, M., Serban, D., and Prusiner, S.B. (1990a). Acquisition of protease resistance by prion proteins in scrapie-infected cells does not require asparagine-linked glycosylation. Proc. Natl. Acad. Sci. U.S.A. *87*, 8262–8266.

167) Taraboulos, A., Serban, D., and Prusiner, S.B. (1990b). Scrapie prion proteins accumulate in the cytoplasm of persistently infected cultured cells. J. Cell Biol. *110*, 2117–2132.

168) Westaway, D., and Prusiner, S.B. (1990). Infectious and genetic manifestations of prion diseases: implications for the BSE epidemic. Nature *346*, 113.

169) Yost, C.S., Lopez, C.D., Prusiner, S.B., Myers, R.M., and Lingappa, V.R. (1990). Non-hydrophobic extracytoplasmic determinant of stop transfer in the prion protein. Nature *343*, 669–672.

170) Carlson, G.A., Hsiao, K., Oesch, B., Westaway, D., and Prusiner, S.B. (1991). Genetics of prion infections. Trends Genet. *7*, 61–65.

171) Epstein, C.J., Foster, D.R., DeArmond, S.J., and Prusiner, S.B. (1991). Acceleration of scrapie in trisomy 16´diploid aggregation chimeras. Ann. Neurol. *29*, 95–97.

172) Hsiao, K., Meiner, Z., Kahana, E., Cass, C., Kahana, I., Avrahami, D., Scarlato, G., Abramsky, O., Prusiner, S.B., and Gabizon, R. (1991a). Mutation of the prion protein in Libyan Jews with Creutzfeldt-Jakob disease. N. Engl. J. Med. *324*, 1091–1097.

173) Hsiao, K., and Prusiner, S.B. (1991). Molecular genetics and transgenic model of Gerstmann-Straussler-Scheinker disease. Alzheimer Dis. Assoc. Disord. *5*, 155–162.

174) Hsiao, K., Scott, M., Foster, D., DeArmond, S.J., Groth, D., Serban, H., and Prusiner, S.B. (1991b). Spontaneous neurodegeneration in transgenic mice with prion protein codon 101 proline->leucine substitution. Ann. N. Y. Acad. Sci. *640*, 166–170.

175) Hsiao, K.K., Cass, C., Schellenberg, G.D., Bird, T., Devine-Gage, E., Wisniewski, H., and Prusiner, S.B. (1991c). A prion protein variant in a family with the telencephalic form of Gerstmann-Sträussler-Scheinker syndrome. Neurology *41*, 681–684.

176) Jendroska, K., Heinzel, F.P., Torchia, M., Stowring, L., Kretzschmar, H.A., Kon, A., Stern, A., Prusiner, S.B., and DeArmond, S.J. (1991). Proteinase-resistant prion protein accumulation in Syrian hamster brain correlates with regional pathology and scrapie infectivity. Neurology *41*, 1482–1490.

177) McKinley, M.P., Meyer, R.K., Kenaga, L., Rahbar, F., Cotter, R., Serban, A., and Prusiner, S.B. (1991a). Scrapie prion rod formation in vitro requires both detergent extraction and limited proteolysis. J. Virol. *65*, 1340–1351.

178) McKinley, M.P., Taraboulos, A., Kenaga, L., Serban, D., Stieber, A., DeArmond, S.J., Prusiner, S.B., and Gonatas, N. (1991b). Ultrastructural localization of scrapie prion proteins in cytoplasmic vesicles of infected cultured cells. Lab. Invest. *65*, 622–630.

179) Meyer, N., Rosenbaum, V., Schmidt, B., Gilles, K., Mirenda, C., Groth, D., Prusiner, S.B., and Riesner, D. (1991). Search for a putative scrapie genome in purified prion fractions reveals a paucity of nucleic acids. J. Gen. Virol. *72*, 37–49.

180) Oesch, B., Westaway, D., and Prusiner, S.B. (1991). Prion protein genes: evolutionary and functional aspects. Curr. Top. Microbiol. Immunol. *172*, 109–124.

181) Prusiner, S.B. (1991a). Molecular biology of prion diseases. Science *252*, 1515–1522.

182) Prusiner, S.B. (1991b). Scrapie prions: molecular genetics and cell biology. Semin. Virol. *2*, 165–179.

183) Prusiner, S.B., and DeArmond, S.J. (1991). Molecular biology and pathology of scrapie and the prion diseases of humans. Brain Pathol. *1*, 297–310.

184) Prusiner, S.B., Torchia, M., and Westaway, D. (1991). Molecular biology and genetics of prions - implications for sheep scrapie, "mad cows" and the BSE epidemic. Cornell Vet. *81*, 85–101.

185) Rogers, M., Serban, D., Gyuris, T., Scott, M., Torchia, T., and Prusiner, S.B. (1991). Epitope mapping of the Syrian hamster prion protein utilizing chimeric and mutant genes in a vaccinia virus expression system. J. Immunol. *147*, 3568–3574.

186) Stahl, N., Baldwin, M.A., and Prusiner, S.B. (1991). Electrospray mass spectrometry of the glycosylinositol phospholipid of the scrapie prion protein. Cell Biol. Int. Rep. *15*, 853–862.

187) Stahl, N., and Prusiner, S.B. (1991). Prions and prion proteins. FASEB J. *5*, 2799–2807.

188) Tagliavini, F., Prelli, F., Ghiso, J., Bugiani, O., Serban, D., Prusiner, S.B., Farlow, M.R., Ghetti, B., and Frangione, B. (1991). Amyloid protein of Gerstmann-Sträussler-Scheinker disease (Indiana kindred) is an 11 kd fragment of prion protein with an N-terminal glycine at codon 58. EMBO J. *10*, 513–519.

189) Taraboulos, A., Raeber, A., Borchelt, D., McKinley, M.P., and Prusiner, S.B. (1991). Brefeldin A inhibits protease resistant prion protein synthesis in scrapie-infected cultured cells. FASEB J. *5*, A1177.

190) Westaway, D., Mirenda, C. A., Foster, D., Zebarjadian, Y., Scott, M., Torchia, M., Yang, S.-L., Serban, H., DeArmond, S. J., Ebeling, C., Prusiner, S. B., and Carlson, G. A. (1991). Paradoxical shortening of scrapie incubation times by expression of prion protein transgenes derived from long incubation period mice. Neuron *7*, 59–68.

191) Borchelt, D.R., Taraboulos, A., and Prusiner, S.B. (1992). Evidence for synthesis of scrapie prion proteins in the endocytic pathway. J. Biol. Chem. *267*, 16188–16199.

192) Büeler, H., Fisher, M., Lang, Y., Bluethmann, H., Lipp, H.-P., DeArmond, S.J., Prusiner, S.B., Aguet, M., and Weissmann, C. (1992). Normal development and behaviour of mice lacking the neuronal cell-surface PrP protein. Nature *356*, 577–582.

193) Dlouhy, S.R., Hsiao, K., Farlow, M.R., Foroud, T., Conneally, P.M., Johnson, P., Prusiner, S.B., Hodes, M.E., and Ghetti, B. (1992). Linkage of the Indiana kindred of Gerstmann-Sträussler-Scheinker disease to the prion protein gene. Nat. Genet. *1*, 64–67.

194) Gabriel, J.-M., Oesch, B., Kretzschmar, H., Scott, M., and Prusiner, S.B. (1992). Molecular cloning of a candidate chicken prion protein. Proc. Natl. Acad. Sci. U.S.A. *89*, 9097–9101.

195) Gasset, M., Baldwin, M.A., Lloyd, D., Gabriel, J.-M., Holtzman, D.M., Cohen, F., Fletterick, R., and Prusiner, S.B. (1992). Predicted a-helical regions of the prion protein when synthesized as peptides form amyloid. Proc. Natl. Acad. Sci. U.S.A. *89*, 10940–10944.

196) Giaccone, G., Verga, L., Bugiani, O., Frangione, B., Serban, D., Prusiner, S.B., Farlow, M.R., Ghetti, B., and Tagliavini, F. (1992). Prion protein preamyloid and amyloid deposits in Gerstmann-Sträussler-Scheinker disease, Indiana kindred. Proc. Natl. Acad. Sci. U.S.A. *89*, 9349–9353.

197) Hecker, R., Taraboulos, A., Scott, M., Pan, K.-M., Torchia, M., Jendroska, K., DeArmond, S.J., and Prusiner, S.B. (1992). Replication of distinct scrapie prion isolates is region specific in brains of transgenic mice and hamsters. Genes Dev. *6*, 1213–1228.

198) Hsiao, K., Dlouhy, S., Farlow, M.R., Cass, C., Da Costa, M., Conneally, M., Hodes, M.E., Ghetti, B., and Prusiner, S.B. (1992). Mutant prion proteins in Gerstmann-Sträussler-Scheinker disease with neurofibrillary tangles. Nat. Genet. *1*, 68–71.

199) Kellings, K., Meyer, N., Mirenda, C., Prusiner, S.B., and Riesner, D. (1992). Further analysis of nucleic acids in purified scrapie prion preparations by improved return refocussing gel electrophoresis (RRGE). J. Gen. Virol. *73*, 1025–1029.

200) Kretzschmar, H.A., Kufer, P., Riethmüller, G., DeArmond, S.J., Prusiner, S.B., and Schiffer, D. (1992a). Prion protein mutation at codon 102 in an Italian family with Gerstmann-Sträussler-Scheinker syndrome. Neurology *42*, 809–810.

201) Kretzschmar, H.A., Neumann, M., Riethmüller, G., and Prusiner, S.B. (1992b). Molecular cloning of a mink prion protein gene. J. Gen. Virol. *73*, 2757–2761.

202) Meiner, Z., Halimi, M., Polakiewicz, R.D., Prusiner, S.B., and Gabizon, R. (1992). Presence of the prion protein in peripheral tissues of Libyan Jews with Creutzfeldt-Jakob disease. Neurology *42*, 1355–1360.

203) Pan, K.-M., Stahl, N., and Prusiner, S.B. (1992). Purification and properties of the cellular prion protein from Syrian hamster brain. Protein Sci. *1*, 1343–1352.

204) Prusiner, S.B. (1992a). Molecular biology and genetics of neurodegenerative diseases caused by prions. Adv. Virus Res. *41*, 241–280.

205) Prusiner, S.B. (1992b). Natural and experimental prion diseases of humans and animals. Curr. Opin. Neurobiol. *2*, 638–647.

206) Prusiner, S.B. (1992c). Chemistry and biology of prions. Biochemistry *31*, 12278–12288.

207) Raeber, A.J., Borchelt, D.R., Scott, M., and Prusiner, S.B. (1992). Attempts to convert the cellular prion protein into the scrapie isoform in cell-free systems. J. Virol. *66*, 6155–6163.

208) Scott, M.R., Köhler, R., Foster, D., and Prusiner, S.B. (1992). Chimeric prion protein expression in cultured cells and transgenic mice. Protein Sci. *1*, 986–997.

209) Stahl, N., Baldwin, M.A., Hecker, R., Pan, K.-M., Burlingame, A.L., and Prusiner, S.B. (1992). Glycosylinositol phospholipid anchors of the scrapie and cellular prion proteins contain sialic acid. Biochemistry *31*, 5043–5053.

210) Taraboulos, A., Jendroska, K., Serban, D., Yang, S.-L., DeArmond, S.J., and Prusiner, S.B. (1992a). Regional mapping of prion proteins in brains. Proc. Natl. Acad. Sci. U.S.A. *89*, 7620–7624.

211) Taraboulos, A., Raeber, A.J., Borchelt, D.R., Serban, D., and Prusiner, S.B. (1992b). Synthesis and trafficking of prion proteins in cultured cells. Mol. Biol. Cell *3*, 851–863.

212) Borchelt, D.R., Rogers, M., Stahl, N., Telling, G., and Prusiner, S.B. (1993). Release of the cellular prion protein from cultured cells after loss of its glycoinositol phospholipid anchor. Glycobiology *3*, 319–329.

213) Carlson, G.A., Ebeling, C., Torchia, M., Westaway, D., and Prusiner, S.B. (1993). Delimiting the location of the scrapie prion incubation time gene on chromosome 2 of the mouse. Genetics *133*, 979–988.

214) DeArmond, S.J., and Prusiner, S.B. (1993). The neurochemistry of prion diseases. J. Neurochem. *61*, 1589–1601.

215) DeArmond, S.J., Yang, S.-L., Lee, A., Bowler, R., Taraboulos, A., Groth, D., and Prusiner, S.B. (1993). Three scrapie prion isolates exhibit different accumulation patterns of the prion protein scrapie isoform. Proc. Natl. Acad. Sci. U.S.A. *90*, 6449–6453.

216) Gabizon, R., Rosenmann, H., Meiner, Z., Kahana, I., Kahana, E., Shugart, Y., Ott, J., and Prusiner, S.B. (1993). Mutation and polymorphism of the prion protein gene in Libyan Jews with Creutzfeldt-Jakob disease (CJD). Am. J. Hum. Genet. *53*, 828–835.

217) Gasset, M., Baldwin, M.A., Fletterick, R.J., and Prusiner, S.B. (1993). Perturbation of the secondary structure of the scrapie prion protein under conditions that alter infectivity. Proc. Natl. Acad. Sci. U.S.A. *90*, 1–5.

218) Kellings, K., Meyer, N., Mirenda, C., Prusiner, S.B., and Riesner, D. (1993). Analysis of nucleic acids in purified scrapie prion preparations. Arch. Virol. Suppl. *7*, 215–225.

219) Kristensson, K., Feuerstein, B., Taraboulos, A., Hyun, W.C., Prusiner, S.B., and DeArmond, S.J. (1993). Scrapie prions alter receptor-mediated calcium responses in cultured cells. Neurology *43*, 2335–2341.

220) Pan, K.-M., Baldwin, M., Nguyen, J., Gasset, M., Serban, A., Groth, D., Mehlhorn, I., Huang, Z., Fletterick, R.J., Cohen, F.E., and Prusiner, S.B. (1993). Conversion of a-helices into b-sheets features in the formation of the scrapie prion proteins. Proc. Natl. Acad. Sci. U.S.A. *90*, 10962–10966.

221) Prusiner, S.B. (1993a). Transgenetics and cell biology of prion diseases - investigations of PrP^{Sc} synthesis and diversity. Br. Med. Bull. *49*, 873–912.

222) Prusiner, S.B. (1993b). Genetic and infectious prion diseases. Arch. Neurol. *50*, 1129–1153.

223) Prusiner, S.B. (1993c). Prion encephalopathies of animals and humans. Dev. Biol. Stand. *80*, 31–44.

224) Prusiner, S.B. (1993d). Transgenetic investigations of prion diseases of humans and animals. Philos. Trans. R. Soc. Lond. B Biol. Sci. *339*, 239–254.

225) Prusiner, S. B., Füzi, M., Scott, M., Serban, D., Serban, H., Taraboulos, A., Gabriel, J.-M., Wells, G. A. H., Wilesmith, J. W., Bradley, R., DeArmond, S. J., and Kristensson, K. (1993a). Immunologic and molecular biologic studies of prion proteins in bovine spongiform encephalopathy. J. Infect. Dis. *167*, 602–613.

226) Prusiner, S.B., Groth, D., Serban, A., Koehler, R., Foster, D., Torchia, M., Burton, D., Yang, S.-L., and DeArmond, S.J. (1993b). Ablation of the prion protein (PrP) gene in mice prevents scrapie and facilitates production of anti-PrP antibodies. Proc. Natl. Acad. Sci. U.S.A. *90*, 10608–10612.

227) Prusiner, S.B., Groth, D., Serban, A., Stahl, N., and Gabizon, R. (1993c). Attempts to restore scrapie prion infectivity after exposure to protein denaturants. Proc. Natl. Acad. Sci. U.S.A. *90*, 2793–2797.

228) Rogers, M., Yehiely, F., Scott, M., and Prusiner, S.B. (1993). Conversion of truncated and elongated prion proteins into the scrapie isoform in cultured cells. Proc. Natl. Acad. Sci. U.S.A. *90*, 3182–3186.

229) Scott, M., Groth, D., Foster, D., Torchia, M., Yang, S.-L., DeArmond, S.J., and Prusiner, S.B. (1993). Propagation of prions with artificial properties in transgenic mice expressing chimeric PrP genes. Cell *73*, 979–988.

230) Stahl, N., Baldwin, M.A., Teplow, D.B., Hood, L., Gibson, B.W., Burlingame, A.L., and Prusiner, S.B. (1993). Structural analysis of the scrapie prion protein using mass spectrometry and amino acid sequencing. Biochemistry *32*, 1991–2002.

231) Baldwin, M.A., Pan, K.-M., Nguyen, J., Huang, Z., Groth, D., Serban, A., Gasset, M., Mehlhorn, I., R.J., F., Cohen, F.E., and Prusiner, S.B. (1994). Spectroscopic characterization of conformational differences between PrP^{C} and PrP^{Sc}: an a-helix to b-sheet transition. Philos. Trans. R. Soc. Lond. B Biol. Sci. *343*, 435–441.

232) Carlson, G.A., DeArmond, S.J., Torchia, M., Westaway, D., and Prusiner, S.B. (1994a). Genetics of prion diseases and prion diversity in mice. Philos. Trans. R. Soc. Lond. B Biol. Sci. *343*, 363–369.

233) Carlson, G.A., Ebeling, C., Yang, S.-L., Telling, G., Torchia, M., Groth, D., Westaway, D., DeArmond, S.J., and Prusiner, S.B. (1994b). Prion isolate specified allotypic interactions between the cellular and scrapie prion proteins in congenic and transgenic mice. Proc. Natl. Acad. Sci. U.S.A. *91*, 5690–5694.

234) Cohen, F.E., Pan, K.-M., Huang, Z., Baldwin, M., Fletterick, R.J., and Prusiner, S.B. (1994). Structural clues to prion replication. Science *264*, 530–531.

235) DeArmond, S.J., Yang, S.-L., Cayetano-Canlas, J., Groth, D., and Prusiner, S.B. (1994). The neuropathological phenotype in transgenic mice expressing different prion protein constructs. Philos. Trans. R. Soc. Lond. B Biol. Sci. *343*, 415–423.

236) DeFea, K.A., Nakahara, D.H., Calayag, M.C., Yost, C.S., Mirels, L.F., Prusiner, S.B., and Lingappa, V.R. (1994). Determinants of carboxyl-terminal domain translocation during prion protein biogenesis. J. Biol. Chem. *269*, 16810–16820.

237) Fink, J.K., Peacock, M.L., Warren, J.T., Roses, A.D., and Prusiner, S.B. (1994). Detecting prion protein gene mutations by denaturing gradient gel electrophoresis. Hum. Mutat. *4*, 42–50.

238) Gabizon, R., Rosenman, H., Meiner, Z., Kahana, I., Kahana, E., Shugart, Y., Ott, J., and Prusiner, S.B. (1994). Mutation in codon 200 and polymorphism in codon 129 of the prion protein gene in Libyan Jews with Creutzfeldt-Jakob disease. Philos. Trans. R. Soc. Lond. B Biol. Sci. *343*, 385–390.

239) Gomi, H., Ikeda, T., Kunieda, T., Itohara, S., Prusiner, S.B., and Yamanouchi, K. (1994). Prion protein (PrP) is not involved in the pathogenesis of spongiform encephalopathy in zitter rats. Neurosci. Lett. *166*, 171–174.

240) Hsiao, K.K., Groth, D., Scott, M., Yang, S.-L., Serban, H., Rapp, D., Foster, D., Torchia, M., DeArmond, S.J., and Prusiner, S.B. (1994). Serial transmission in rodents of neurodegeneration from transgenic mice expressing mutant prion protein. Proc. Natl. Acad. Sci. U.S.A. *91*, 9126–9130.

241) Huang, Z., Gabriel, J.-M., Baldwin, M.A., Fletterick, R.J., Prusiner, S.B., and Cohen, F.E. (1994). Proposed three-dimensional structure for the cellular prion protein. Proc. Natl. Acad. Sci. U.S.A. *91*, 7139–7143.

242) Jefferys, J.G.R., Empson, R.M., Whittington, M.A., and Prusiner, S.B. (1994). Scrapie infection of transgenic mice leads to network and intrinsic dysfunction of cortical and hippocampal neurones. Neurobiol. Dis. *1*, 25–30.

243) Kellings, K., Prusiner, S.B., and Riesner, D. (1994). Nucleic acids in prion preparations: unspecific background or essential component? Philos. Trans. R. Soc. Lond. B Biol. Sci. *343*, 425–430.

244) Kolbert, A.C., Grandinetti, P.J., Baldwin, M., Prusiner, S.B., and Pines, A. (1994). Measurement of internuclear distances by switched angle spinning NMR. J. Phys. Chem. *98*, 7936–7938.

245) Prusiner, S.B. (1994a). Inherited prion diseases. Proc. Natl. Acad. Sci. U.S.A. *91*, 4611–4614.

246) Prusiner, S.B. (1994b). Molecular biology and genetics of prion diseases. Philos. Trans. R. Soc. Lond. B Biol. Sci. *343*, 447–463.

247) Prusiner, S.B. (1994c). Biology and genetics of prion diseases. Annu. Rev. Microbiol. *48*, 655–686.

248) Prusiner, S.B. (1994d). A national strategy for development of effective methods for the prevention and treatment of Alzheimer's disease and related neurodegenerative disorders. Neurobiol. Aging *15 Supp2*, S29-S32.

249) Prusiner, S.B., and DeArmond, S.J. (1994). Prion diseases and neurodegeneration. Annu. Rev. Neurosci. *17*, 311–339.

250) Prusiner, S.B., and Hsiao, K.K. (1994). Human prion diseases. Ann. Neurol. *35*, 385–395.

251) Taraboulos, A., Scott, M., Semenov, A., Avrahami, D., and Prusiner, S.B. (1994). Biosynthesis of the prion proteins in scrapie-infected cells in culture. Braz. J. Med. Biol. Res. *27*, 303–307.

252) Telling, G.C., Scott, M., Hsiao, K.K., Foster, D., Yang, S.-L., Torchia, M., Sidle, K.C.L., Collinge, J., DeArmond, S.J., and Prusiner, S.B. (1994). Transmission of Creutzfeldt-Jakob disease from humans to transgenic mice expressing chimeric human-mouse prion protein. Proc. Natl. Acad. Sci. U.S.A. *91*, 9936–9940.

253) Westaway, D., Cooper, C., Turner, S., Da Costa, M., Carlson, G.A., and Prusiner, S.B. (1994a). Structure and polymorphism of the mouse prion protein gene. Proc. Natl. Acad. Sci. U.S.A. *91*, 6418–6422.

254) Westaway, D., DeArmond, S.J., Cayetano-Canlas, J., Groth, D., Foster, D., Yang, S.-L., Torchia, M., Carlson, G.A., and Prusiner, S.B. (1994b). Degeneration of skeletal muscle, peripheral nerves, and the central nervous system in transgenic mice overexpressing wild-type prion proteins. Cell *76*, 117–129.

255) Westaway, D., Zuliani, V., Cooper, C.M., Da Costa, M., Neuman, S., Jenny, A.L., Detwiler, L., and Prusiner, S.B. (1994c). Homozygosity for prion protein alleles encoding glutamine-171 renders sheep susceptible to natural scrapie. Genes Dev. *8*, 959–969.

256) Baldwin, M.A., Cohen, F.E., and Prusiner, S.B. (1995). Prion protein isoforms, a convergence of biological and structural investigations. J. Biol. Chem. *270*, 19197–19200.

257) DeArmond, S.J., and Prusiner, S.B. (1995a). Prion protein transgenes and the neuropathology in prion diseases. Brain Pathol. *5*, 77–89.

258) DeArmond, S.J., and Prusiner, S.B. (1995b). Etiology and pathogenesis of prion diseases. Am. J. Pathol. *146*, 785–811.

259) Huang, Z., Prusiner, S.B., and Cohen, F.E. (1995). Scrapie prions: a three-dimensional model of an infectious fragment. Fold. Des. *1*, 13–19.

260) Kaneko, K., Peretz, D., Pan, K.-M., Blochberger, T., Wille, H., Gabizon, R., Griffith, O.H., Cohen, F.E., Baldwin, M.A., and Prusiner, S.B. (1995). Prion protein (PrP) synthetic peptides induce cellular PrP to acquire properties of the scrapie isoform. Proc. Natl. Acad. Sci. U.S.A. *92*, 11160–11164.

261) Kazmirski, S.L., Alonso, D.O.V., Cohen, F.E., Prusiner, S.B., and Daggett, V. (1995). Theoretical studies of sequence effects on the conformational properties of a fragment of the prion protein: implications for scrapie formation. Chem. Biol. *2*, 305–315.

262) Mastrianni, J.A., Curtis, M.T., Oberholtzer, J.C., Da Costa, M.M., DeArmond, S., Prusiner, S.B., and Garbern, J.Y. (1995). Prion disease (PrP-A117V) presenting with ataxia instead of dementia. Neurology *45*, 2042–2050.

263) Nguyen, J., Baldwin, M.A., Cohen, F.E., and Prusiner, S.B. (1995a). Prion protein peptides induce a-helix to b-sheet conformational transitions. Biochemistry *34*, 4186–4192.

264) Nguyen, J.T., Inouye, H., Baldwin, M.A., Fletterick, R.J., Cohen, F.E., Prusiner, S.B., and Kirschner, D.A. (1995b). X-ray diffraction of scrapie prion rods and PrP peptides. J. Mol. Biol. *252*, 412–422.

265) Prusiner, S.B. (1995). The prion diseases. Sci. Am. *272*, 30–37.

266) Prusiner, S.B., Baldwin, M., Collinge, J., DeArmond, S.J., Marsh, R., Tateishi, J., and Weissmann, C. (1995). Subviral agents: agents of spongiform encephalopathies (prions). Arch. Virol. Suppl. *10*, 498–503.

267) Prusiner, S.B., and DeArmond, S.J. (1995). Prion protein amyloid and neurodegeneration. Amyloid: Int. J. Exp. Clin. Invest. *2*, 39–65.

268) Raeber, A.J., Muramoto, T., Kornberg, T.B., and Prusiner, S.B. (1995). Expression and targeting of Syrian hamster prion protein induced by heat shock in transgenic *Drosophila melanogaster*. Mech. Dev. *51*, 317–327.

269) Schätzl, H.M., Da Costa, M., Taylor, L., Cohen, F.E., and Prusiner, S.B. (1995). Prion protein gene variation among primates. J. Mol. Biol. *245*, 362–374.

270) Spudich, S., Mastrianni, J.A., Wrensch, M., Gabizon, R., Meiner, Z., Kahana, I., Rosenmann, H., Kahana, E., and Prusiner, S.B. (1995). Complete penetrance of Creutzfeldt-Jakob disease in Libyan Jews carrying the E200K mutation in the prion protein gene. Mol. Med. *1*, 607–613.

271) Taraboulos, A., Scott, M., Semenov, A., Avrahami, D., Laszlo, L., and Prusiner, S.B. (1995). Cholesterol depletion and modification of COOH-terminal targeting sequence of the prion protein inhibits formation of the scrapie isoform. J. Cell Biol. *129*, 121–132.

272) Tatzelt, J., Zuo, J., Voellmy, R., Scott, M., Hartl, U., Prusiner, S.B., and Welch, W.J. (1995). Scrapie prions selectively modify the stress response in neuroblastoma cells. Proc. Natl. Acad. Sci. U.S.A. *92*, 2944–2948.

273) Telling, G.C., Scott, M., Mastrianni, J., Gabizon, R., Torchia, M., Cohen, F.E., DeArmond, S.J., and Prusiner, S.B. (1995). Prion propagation in mice expressing human and chimeric PrP transgenes implicates the interaction of cellular PrP with another protein. Cell *83*, 79–90.

274) Westaway, D., Carlson, G.A., and Prusiner, S.B. (1995). On safari with PrP: prion diseases of animals. Trends Microbiol. *3*, 141–147.

275) Wiese, U., Wulfert, M., Prusiner, S.B., and Riesner, D. (1995). Scanning for mutations in the human prion protein open reading frame by temporal temperature gradient gel electrophoresis. Electrophoresis *16*, 1851–1860.

276) Zhang, H., Kaneko, K., Nguyen, J.T., Livshits, T.L., Baldwin, M.A., Cohen, F.E., James, T.L., and Prusiner, S.B. (1995). Conformational transitions in peptides containing two putative a-helices of the prion protein. J. Mol. Biol. *250*, 514–526.

277) Bamborough, P., Wille, H., Telling, G.C., Yehiely, F., Prusiner, S.B., and Cohen, F.E. (1996). Prion protein structure and scrapie replication: theoretical, spectroscopic and genetic investigations. Cold Spring Harb. Symp. Quant. Biol. *61*, 495–509.

278) DeArmond, S.J., Qiu, Y., Wong, K., Nixon, R., Hyun, W., Prusiner, S.B., and Mobley, W.C. (1996). Abnormal plasma membrane properties and functions in prion-infected cell lines. Cold Spring Harb. Symp. Quant. Biol. *61*, 531–540.

279) Diez, M., Koistinaho, J., DeArmond, S.J., Camerino, A.P., Groth, D., Caytano, J.C., Prusiner, S.B., and Hökfelt, T. (1996). Aberrant induction of neuropeptide Y mRNA in hippocampal CA3 pyramidal neurones in scrapie-infected mice. Neuroreport *7*, 1887–1892.

280) Gabizon, R., Telling, G., Meiner, Z., Halimi, M., Kahana, I., and Prusiner, S.B. (1996). Insoluble wild-type and protease-resistant mutant prion protein in brains of patients with inherited prion disease. Nat. Med. *2*, 59–64.

281) Guan, Z., Söderberg, M., Sindelar, P., Prusiner, S.B., Kristensson, K., and Dallner, G. (1996). Lipid composition in scrapie-infected mouse brain: prion infection increases the levels of dolichyl phosphate and ubiquinone. J. Neurochem. *66*, 277–285.

282) Heller, J., Kolbert, A.C., Larsen, R., Ernst, M., Bekker, T., Baldwin, M., Prusiner, S.B., Pines, A., and Wemmer, D.E. (1996a). Solid-state NMR studies of the prion protein H1 fragment. Protein Sci. *5*, 1655–1661.

283) Heller, J., Larsen, R., Ernst, M., Kolbert, A.C., Baldwin, M., Prusiner, S.B., Wemmer, D.E., and Pines, A. (1996b). Application of rotational resonance to inhomogeneously broadened systems. Chem. Phys. Lett. *251*, 223–229.

284) Lledo, P.-M., Tremblay, P., DeArmond, S.J., Prusiner, S.B., and Nicoll, R.A. (1996). Mice deficient for prion protein exhibit normal neuronal excitability and synaptic transmission in the hippocampus. Proc. Natl. Acad. Sci. U.S.A. *93*, 2403–2407.

285) Mastrianni, J.A., Iannicola, C., Myers, R.M., DeArmond, S., and Prusiner, S.B. (1996). Mutation of the prion protein gene at codon 208 in familial Creutzfeldt- Jakob disease. Neurology *47*, 1305–1312.

286) Mehlhorn, I., Groth, D., Stöckel, J., Moffat, B., Reilly, D., Yansura, D., Willett, W. S., Baldwin, M., Fletterick, R., Cohen, F. E., Vandlen, R., Henner, D., and Prusiner, S. B. (1996). High-level expression and characterization of a purified 142-residue polypeptide of the prion protein. Biochemistry *35*, 5528–5537.

287) Muramoto, T., Scott, M., Cohen, F.E., and Prusiner, S.B. (1996). Recombinant scrapie-like prion protein of 106 amino acids is soluble. Proc. Natl. Acad. Sci. U.S.A. *93*, 15457–15462.

288) Prusiner, S.B. (1996a). Prion biology and diseases-laughing cannibals, mad cows, and scientific heresy. Med. Res. Rev. *16*, 487–505.

289) Prusiner, S.B. (1996b). Molecular biology and pathogenesis of prion diseases. Trends Biochem. Sci. *252*, 482–487.

290) Prusiner, S.B. (1996c). Molecular biology and genetics of prion diseases. Cold Spring Harb. Symp. Quant. Biol. *61*, 473–493.

291) Riesner, D., Kellings, K., Post, K., Wille, H., Serban, H., Groth, D., Baldwin, M.A., and Prusiner, S.B. (1996). Disruption of prion rods generates 10-nm spherical particles having high a-helical content and lacking scrapie infectivity. J. Virol. *70*, 1714–1722.

292) Tatzelt, J., Maeda, N., Pekny, M., Yang, S.-L., Betsholtz, C., Eliasson, C., Cayetano, J., Camerino, A.P., DeArmond, S.J., and Prusiner, S.B. (1996a). Scrapie in mice deficient in apolipoprotein E or glial fibrillary acidic protein. Neurology *47*, 449–453.

293) Tatzelt, J., Prusiner, S.B., and Welch, W.J. (1996b). Chemical chaperones interfere with the formation of scrapie prion protein. EMBO J. *15*, 6363–6373.

294) Telling, G.C., Haga, T., Torchia, M., Tremblay, P., DeArmond, S.J., and Prusiner, S.B. (1996a). Interactions between wild-type and mutant prion proteins modulate neurodegeneration in transgenic mice. Genes Dev. *10*, 1736–1750.

295) Telling, G.C., Parchi, P., DeArmond, S.J., Cortelli, P., Montagna, P., Gabizon, R., Mastrianni, J., Lugaresi, E., Gambetti, P., and Prusiner, S.B. (1996b). Evidence for the conformation of the pathologic isoform of the prion protein enciphering and propagating prion diversity. Science *274*, 2079–2082.

296) Vey, M., Pilkuhn, S., Wille, H., Nixon, R., DeArmond, S.J., Smart, E.J., Anderson, R.G., Taraboulos, A., and Prusiner, S.B. (1996). Subcellular colocalization of the cellular and scrapie prion proteins in caveolae-like membranous domains. Proc. Natl. Acad. Sci. U.S.A. *93*, 14945–14949.

297) Wille, H., Zhang, G.-F., Baldwin, M.A., Cohen, F.E., and Prusiner, S.B. (1996). Separation of scrapie prion infectivity from PrP amyloid polymers. J. Mol. Biol. *259*, 608–621.

298) Williamson, R.A., Peretz, D., Smorodinsky, N., Bastidas, R., Serban, H., Mehlhorn, I., DeArmond, S.J., Prusiner, S.B., and Burton, D.R. (1996). Circumventing tolerance to generate autologous monoclonal antibodies to the prion protein. Proc. Natl. Acad. Sci. U.S.A. *93*, 7279–7282.

299) Wong, K., Qiu, Y., Hyun, W., Nixon, R., VanCleff, J., Sanchez-Salazar, J., Prusiner, S.B., and DeArmond, S.J. (1996). Decreased receptor-mediated calcium response in prion-infected cells correlates with decreased membrane fluidity and IP_3 release. Neurology *47*, 741–750.

300) Blochberger, T.C., Cooper, C., Peretz, D., Tatzelt, J., Griffith, O.H., Baldwin, M.A., and Prusiner, S.B. (1997). Prion protein expression in Chinese hamster ovary cells using a glutamine synthetase selection and amplification system. Protein Eng. *10*, 1465–1473.

301) Bosque, P.J., Telling, G.C., Cayetano, J., DeArmond, S.J., and Prusiner, S.B. (1997). Evidence for prion replication in skeletal muscle. Ann. Neurol. *42*, 986.

302) Carlson, G.A., Banks, S., Lund, D., Reichert, C., Groth, D., Torchia, M., DeArmond, S.J., and Prusiner, S.B. (1997). Failure to transmit disease from gray tremor mutant mice. J. Virol. *71*, 2342–2345.

303) DeArmond, S. J., Sánchez, H., Yehiely, F., Qiu, Y., Ninchak-Casey, A., Daggett, V., Camerino, A. P., Cayetano, J., Rogers, M., Groth, D., Torchia, M., Tremblay, P., Scott, M. R., Cohen, F. E., and Prusiner, S. B. (1997). Selective neuronal targeting in prion disease. Neuron *19*, 1337–1348.

304) Diez, M., Koistinaho, J., DeArmond, S.J., Groth, D., Prusiner, S.B., and Hökfelt, T. (1997). Marked decrease of neuropeptide Y Y2 receptor binding sites in the hippocampus in murine prion disease. Proc. Natl. Acad. Sci. U.S.A. *94*, 13267–13272.

305) Donne, D.G., Viles, J.H., Groth, D., Mehlhorn, I., James, T.L., Cohen, F.E., Prusiner, S.B., Wright, P.E., and Dyson, H.J. (1997). Structure of the recombinant full-length hamster prion protein PrP(29–231): the N terminus is highly flexible. Proc. Natl. Acad. Sci. U.S.A. *94*, 13452–13457.

306) Harrison, P.M., Bamborough, P., Daggett, V., Prusiner, S.B., and Cohen, F.E. (1997). The prion folding problem. Curr. Opin. Struct. Biol. *7*, 53–59.

307) James, T.L., Liu, H., Ulyanov, N.B., Farr-Jones, S., Zhang, H., Donne, D.G., Kaneko, K., Groth, D., Mehlhorn, I., Prusiner, S.B., and Cohen, F.E. (1997). Solution structure of a 142-residue recombinant prion protein corresponding to the infectious fragment of the scrapie isoform. Proc. Natl. Acad. Sci. U.S.A. *94*, 10086–10091.

308) Kaneko, K., Vey, M., Scott, M., Pilkuhn, S., Cohen, F.E., and Prusiner, S.B. (1997a). COOH-terminal sequence of the cellular prion protein directs subcellular trafficking and controls conversion into the scrapie isoform. Proc. Natl. Acad. Sci. U.S.A. *94*, 2333–2338.

309) Kaneko, K., Wille, H., Mehlhorn, I., Zhang, H., Ball, H., Cohen, F.E., Baldwin, M.A., and Prusiner, S.B. (1997b). Molecular properties of complexes formed between the prion protein and synthetic peptides. J. Mol. Biol. *270*, 574–586.

310) Kaneko, K., Zulianello, L., Scott, M., Cooper, C.M., Wallace, A.C., James, T.L., Cohen, F.E., and Prusiner, S.B. (1997c). Evidence for protein X binding to a discontinuous epitope on the cellular prion protein during scrapie prion propagation. Proc. Natl. Acad. Sci. U.S.A. *94*, 10069–10074.

311) Lundberg, K.M., Stenland, C.J., Cohen, F.E., Prusiner, S.B., and Millhauser, G.L. (1997). Kinetics and mechanism of amyloid formation by the prion protein H1 peptide as determined by time-dependent ESR. Chem. Biol. *4*, 345–355.

312) Mastrianni, J., Nixon, F., Layzer, R., DeArmond, S.J., and Prusiner, S.B. (1997). Fatal sporadic insomnia: fatal familial insomnia phenotype without a mutation of the prion protein gene. Neurology *48 [Suppl.]*, A296.

313) Meiner, Z., Gabizon, R., and Prusiner, S.B. (1997). Familial Creutzfeldt-Jakob disease—Codon 200 prion disease in Libyan Jews. Medicine *76*, 227–237.

314) Muramoto, T., DeArmond, S.J., Scott, M., Telling, G.C., Cohen, F.E., and Prusiner, S.B. (1997). Heritable disorder resembling neuronal storage disease in mice expressing prion protein with deletion of an a-helix. Nat. Med. *3*, 750–755.

315) Peretz, D., Williamson, R. A., Matsunaga, Y., Serban, H., Pinilla, C., Bastidas, R. B., Rozenshteyn, R., James, T. L., Houghten, R. A., Cohen, F. E., Prusiner, S. B., and Burton, D. R. (1997). A conformational transition at the N-terminus of the prion protein features in formation of the scrapie isoform. J. Mol. Biol. *273*, 614–622.

316) Prusiner, S.B. (1997). Prion diseases and the BSE crisis. Science *278*, 245–251.

317) Prusiner, S.B., and Scott, M.R. (1997). Genetics of prions. Annu. Rev. Genet. *31*, 139–175.

318) Schätzl, H.M., Da Costa, M., Taylor, L., Cohen, F.E., and Prusiner, S.B. (1997a). Prion protein gene variation among primates (Corrigendum). J. Mol. Biol. *265*, 257.

319) Schätzl, H.M., Laszlo, L., Holtzman, D.M., Tatzelt, J., DeArmond, S.J., Weiner, R.I., Mobley, W.C., and Prusiner, S.B. (1997b). A hypothalamic neuronal cell line persistently infected with scrapie prions exhibits apoptosis. J. Virol. *71*, 8821–8831.

320) Scott, M.R., Groth, D., Tatzelt, J., Torchia, M., Tremblay, P., DeArmond, S.J., and Prusiner, S.B. (1997a). Propagation of prion strains through specific conformers of the prion protein. J. Virol. *71*, 9032–9044.

321) Scott, M. R., Safar, J., Telling, G., Nguyen, O., Groth, D., Torchia, M., Koehler, R., Tremblay, P., Walther, D., Cohen, F. E., DeArmond, S. J., and Prusiner, S. B. (1997b). Identification of a prion protein epitope modulating transmission of bovine spongiform encephalopathy prions to transgenic mice. Proc. Natl. Acad. Sci. U.S.A. *94*, 14279–14284.

322) Telling, G.C., Tremblay, P., Torchia, M., DeArmond, S.J., Cohen, F.E., and Prusiner, S.B. (1997). N-terminally tagged prion protein supports prion propagation in transgenic mice. Protein Sci. *6*, 825–833.

323) Yehiely, F., Bamborough, P., Costa, M.D., Perry, B.J., Thinakaran, G., Cohen, F.E., Carlson, G.A., and Prusiner, S.B. (1997). Identification of candidate proteins binding to prion protein. Neurobiol. Dis. *3*, 339–355.

324) Zhang, H., Stöckel, J., Mehlhorn, I., Groth, D., Baldwin, M.A., Prusiner, S.B., James, T.L., and Cohen, F.E. (1997). Physical studies of conformational plasticity in a recombinant prion protein. Biochemistry *36*, 3543–3553.

325) Cohen, F.E., and Prusiner, S.B. (1998). Pathologic conformations of prion proteins. Annu. Rev. Biochem. *67*, 793–819.

326) Hegde, R.S., Mastrianni, J.A., Scott, M.R., DeFea, K.A., Tremblay, P., Torchia, M., DeArmond, S.J., Prusiner, S.B., and Lingappa, V.R. (1998). A transmembrane form of the prion protein in neurodegenerative disease. Science *279*, 827–834.

327) Hökfelt, T., Broberger, C., Zhang, X., Diez, M., Kopp, J., Xu, Z.-Q., Landry, M., Bao, L., Schalling, M., Koistinaho, J., DeArmond, S. J., Prusiner, S., Gong, J., and Walsh, J. H. (1998). Neuropeptide Y: some viewpoints on a multifaceted peptide in the normal and diseased nervous system. Brain Res. Brain Res. Rev. *26*, 154–166.

328) Lee, I. Y., Westaway, D., Smit, A. F. A., Wang, K., Seto, J., Chen, L., Acharya, C., Ankener, M., Baskin, D., Cooper, C., Yao, H., Prusiner, S. B., and Hood, L. E. (1998). Complete genomic sequence and analysis of the prion protein gene region from three mammalian species. Genome Res. *8*, 1022–1037.

329) Liu, H., Farr-Jones, S., Ulyanov, N., Llinas, M., Marqusee, S., Cohen, F.E., Prusiner, S.B., and James, T.L. (1998). Solution structure of a prion protein: implications for infectivity. J. Kor. Magn. Reson. *2*, 85–105.

330) Post, K., Pitschke, M., Schafer, O., Wille, H., Appel, T.R., Kirsch, D., Mehlhorn, I., Serban, H., Prusiner, S.B., and Riesner, D. (1998). Rapid acquisition of b-sheet structure in the prion protein prior to multimer formation. Biol. Chem. *379*, 1307–1317.

331) Prusiner, S.B. (1998). Prions. Proc. Natl. Acad. Sci. U.S.A. *95*, 13363–13383.

332) Prusiner, S.B., Scott, M.R., DeArmond, S.J., and Cohen, F.E. (1998). Prion protein biology. Cell *93*, 337–348.

333) Safar, J., Wille, H., Itri, V., Groth, D., Serban, H., Torchia, M., Cohen, F.E., and Prusiner, S.B. (1998). Eight prion strains have PrP^{Sc} molecules with different conformations. Nat. Med. *4*, 1157–1165.

334) Stöckel, J., Safar, J., Wallace, A.C., Cohen, F.E., and Prusiner, S.B. (1998). Prion protein selectively binds copper (II) ions. Biochemistry *37*, 7185–7193.

335) Tremblay, P., Meiner, Z., Galou, M., Heinrich, C., Petromilli, C., Lisse, T., Cayetano, J., Torchia, M., Mobley, W., Bujard, H., DeArmond, S. J., and Prusiner, S. B. (1998). Doxycyline control of prion protein transgene expression modulates prion disease in mice. Proc. Natl. Acad. Sci. U.S.A. *95*, 12580–12585.

336) Williamson, R.A., Peretz, D., Pinilla, C., Ball, H., Bastidas, R.B., Rozenshteyn, R., Houghten, R.A., Prusiner, S.B., and Burton, D.R. (1998). Mapping the prion protein using recombinant antibodies. J. Virol. *72*, 9413–9418.

337) Harrison, P.M., Chan, H.S., Prusiner, S.B., and Cohen, F.E. (1999). Thermodynamics of model prions and its implications for the problem of prion protein folding. J. Mol. Biol. *286*, 593–606.

338) Hegde, R.S., Tremblay, P., Groth, D., Prusiner, S.B., and Lingappa, V.R. (1999). Transmissible and genetic prion diseases share a common pathway of neurodegeneration. Nature *402*, 822–826.

339) Kanyo, Z.F., Pan, K.-M., Williamson, A., Burton, D.R., Prusiner, S.B., Fletterick, R.J., and Cohen, F.E. (1999). Antibody binding defines a structure for an epitope that participates in the PrP^{C} -> PrP^{Sc} conformational change. J. Mol. Biol. *293*, 855–863.

340) Liu, H., Farr-Jones, S., Ulyanov, N.B., Llinas, M., Marqusee, S., Groth, D., Cohen, F.E., Prusiner, S.B., and James, T.L. (1999). Solution structure of Syrian hamster prion protein rPrP(90–231). Biochemistry *38*, 5362–5377.

341) Mastrianni, J.A., Nixon, R., Layzer, R., Telling, G.C., Han, D., DeArmond, S.J., and Prusiner, S.B. (1999). Prion protein conformation in a patient with sporadic fatal insomnia. N. Engl. J. Med. *340*, 1630–1638.

342) Moore, R. C., Lee, I. Y., Silverman, G. L., Harrison, P. M., Strome, R., Heinrich, C., Karunaratne, A., Pasternak, S. H., Chishti, M. A., Liang, Y., Mastrangelo, P., Wang, K., Smit, A. F. A., Katamine, S., Carlson, G. A., Cohen, F. E., Prusiner, S. B., Melton, D. W., Tremblay, P., Hood, L. E., and Westaway, D. (1999). Ataxia in prion protein (PrP)-deficient mice is associated with upregulation of the novel PrP-like protein doppel. J. Mol. Biol. *292*, 797–817.

343) Nishida, N., Tremblay, P., Sugimoto, T., Shigematsu, K., Shirabe, S., Petromilli, C., Erpel, S. P., Nakaoke, R., Atarashi, R., Houtani, T., Torchia, M., Sakaguchi, S., DeArmond, S. J., Prusiner, S. B., and Katamine, S. (1999). A mouse prion protein transgene rescues mice deficient for the prion protein gene from Purkinje cell degeneration and demyelination. Lab. Invest. *79*, 689–697.

344) Rudd, P.M., Endo, T., Colominas, C., Groth, D., Wheeler, S.F., Harvey, D.J., Wormald, M.R., Serban, H., Prusiner, S.B., Kobata, A., and Dwek, R.A. (1999). Glycosylation differences between the normal and pathogenic prion protein isoforms. Proc. Natl. Acad. Sci. U.S.A. *96*, 13044–13049.

345) Safar, J., Prusiner, S.B., and DeArmond, S.J. (1999). Diagnosis and pathogenesis of prion diseases. Biomed. Prog. *12*, 27–33.

346) Scott, M.R., Will, R., Ironside, J., Nguyen, H.-O.B., Tremblay, P., DeArmond, S.J., and Prusiner, S.B. (1999). Compelling transgenetic evidence for transmission of bovine spongiform encephalopathy prions to humans. Proc. Natl. Acad. Sci. U.S.A. *96*, 15137–15142.

347) Supattapone, S., Bosque, P., Muramoto, T., Wille, H., Aagaard, C., Peretz, D., Nguyen, H.-O. B., Heinrich, C., Torchia, M., Safar, J., Cohen, F. E., DeArmond, S. J., Prusiner, S. B., and Scott, M. (1999a). Prion protein of 106 residues creates an artificial transmission barrier for prion replication in transgenic mice. Cell *96*, 869–878.

348) Supattapone, S., Nguyen, H.-O.B., Cohen, F.E., Prusiner, S.B., and Scott, M.R. (1999b). Elimination of prions by branched polyamines and implications for therapeutics. Proc. Natl. Acad. Sci. U.S.A. *96*, 14529–14534.

349) Tatzelt, J., Groth, D.F., Torchia, M., Prusiner, S.B., and DeArmond, S.J. (1999). Kinetics of prion protein accumulation in the CNS of mice with experimental scrapie. J. Neuropathol. Exp. Neurol. *58*, 1244–1249.

350) Viles, J.H., Cohen, F.E., Prusiner, S.B., Goodin, D.B., Wright, P.E., and Dyson, H.J. (1999). Copper binding to the prion protein: structural implications of four identical cooperative binding sites. Proc. Natl. Acad. Sci. U.S.A. *96*, 2042–2047.

351) Wille, H., and Prusiner, S.B. (1999). Ultrastructural studies on scrapie prion protein crystals obtained from reverse micellar solutions. Biophys. J. *76*, 1048–1062.

352) Aronoff-Spencer, E., Burns, C.S., Avdievich, N.I., Gerfen, G.J., Peisach, J., Antholine, W.E., Ball, H.L., Cohen, F.E., Prusiner, S.B., and Millhauser, G.L. (2000). Identification of the Cu^{2+} binding sites in the N-terminal domain of the prion protein by EPR and CD spectroscopy. Biochemistry *39*, 13760–13771.

353) Baskakov, I.V., Aagaard, C., Mehlhorn, I., Wille, H., Groth, D., Baldwin, M.A., Prusiner, S.B., and Cohen, F.E. (2000). Self-assembly of recombinant prion protein of 106 residues. Biochemistry *39*, 2792–2804.

354) Bosque, P.J., and Prusiner, S.B. (2000). Cultured cell sublines highly susceptible to prion infection. J. Virol. *74*, 4377–4386.

355) Bouzamondo, E., Milroy, A.M., Ralston, H.J., Prusiner, S.B., and DeArmond, S.J. (2000). Selective neuronal vulnerability during experimental scrapie infection: Insights from an ultrastructural investigation. Brain Res. *874*, 210–215.

356) Inouye, H., Bond, J., Baldwin, M.A., Ball, H.L., Prusiner, S.B., and Kirschner, D.A. (2000). Structural changes in a hydrophobic domain of the prion protein induced by hydration and by Ala -> Val and Pro -> Leu substitutions. J. Mol. Biol. *300*, 1283–1296.

357) Kaneko, K., Ball, H. L., Wille, H., Zhang, H., Groth, D., Torchia, M., Tremblay, P., Safar, J., Prusiner, S. B., DeArmond, S. J., Baldwin, M. A., and Cohen, F. E. (2000). A synthetic peptide initiates Gerstmann-Sträussler-Scheinker (GSS) disease in transgenic mice. J. Mol. Biol. *295*, 997–1007.

358) Korth, C., Kaneko, K., and Prusiner, S.B. (2000). Expression of unglycosylated mutated prion protein facilitates PrP^{Sc} formation in neuroblastoma cells infected with different prion strains. J. Gen. Virol. *81*, 2555–2563.

359) Perrier, V., Wallace, A.C., Kaneko, K., Safar, J., Prusiner, S.B., and Cohen, F.E. (2000). Mimicking dominant negative inhibition of prion replication through structure-based drug design. Proc. Natl. Acad. Sci. U.S.A. *97*, 6073–6078.

360) Schlumpberger, M., Wille, H., Baldwin, M.A., Butler, D.A., Herskowitz, I., and Prusiner, S.B. (2000). The prion domain of yeast Ure2p induces autocatalytic formation of amyloid fibers by a recombinant fusion protein. Protein Sci. *9*, 440–451.

361) Silverman, G.L., Qin, K., Moore, R.C., Yang, Y., Mastrangelo, P., Tremblay, P., Prusiner, S.B., Cohen, F.E., and Westaway, D. (2000). Doppel is an *N*-glycosylated, glycosylphosphatidylinositol-anchored protein. J. Biol. Chem. *275*, 26834–26841.

362) Stephenson, D.A., Chiotti, K., Ebeling, C., Groth, D., DeArmond, S.J., Prusiner, S.B., and Carlson, G.A. (2000). Quantitative trait loci affecting prion incubation time in mice. Genomics *69*, 47–53.

363) Supattapone, S., Nguyen, H.-O.B., Muramoto, T., Cohen, F.E., DeArmond, S.J., Prusiner, S.B., and Scott, M. (2000). Affinity-tagged miniprion derivatives spontaneously adopt protease-resistant conformations. J. Virol. *74*, 11928–11934.

364) Whittal, R.M., Ball, H.L., Cohen, F.E., Burlingame, A.L., Prusiner, S.B., and Baldwin, M.A. (2000). Copper binding to octarepeat peptides of the prion protein monitored by mass spectrometry. Protein Sci. *9*, 332–343.

365) Wille, H., Prusiner, S.B., and Cohen, F.E. (2000). Scrapie infectivity is independent of amyloid staining properties of the N-terminally truncated prion protein. J. Struct. Biol. *130*, 323–338.

366) Zulianello, L., Kaneko, K., Scott, M., Erpel, S., Han, D., Cohen, F.E., and Prusiner, S.B. (2000). Dominant-negative inhibition of prion formation diminished by deletion mutagenesis of the prion protein. J. Virol. *74*, 4351–4360.

367) Ball, H.L., King, D.S., Cohen, F.E., Prusiner, S.B., and Baldwin, M.A. (2001). Engineering the prion protein using chemical synthesis. J. Pept. Res. *58*, 357–374.

368) Baskakov, I.V., Legname, G., Prusiner, S.B., and Cohen, F.E. (2001). Folding of prion protein to its native a-helical conformation is under kinetic control. J. Biol. Chem. *276*, 19687–19690.

369) Diez, M., DeArmond, S.J., Groth, D., Prusiner, S.B., and Hökfelt, T. (2001). Decreased MK-801 binding in discrete hippocampal regions of prion-infected mice. Neurobiol. Dis. *8*, 692–699.

370) Harrison, P.M., Chan, H.S., Prusiner, S.B., and Cohen, F.E. (2001). Conformational propagation with prion-like characteristics in a simple model of protein folding. Protein Sci. *10*, 819–835.

371) Jansen, K., Schäfer, O., Birkmann, E., Post, K., Serban, H., Prusiner, S.B., and Riesner, D. (2001). Structural intermediates in the putative pathway from the cellular prion protein to the pathogenic form. Biol. Chem. *382*, 683–691.

372) Korth, C., May, B.C.H., Cohen, F.E., and Prusiner, S.B. (2001). Acridine and phenothiazine derivatives as pharmacotherapeutics for prion disease. Proc. Natl. Acad. Sci. U.S.A. *98*, 9836–9841.

373) Laws, D.D., Bitter, H.-M.L., Liu, K., Ball, H.L., Kaneko, K., Wille, H., Cohen, F.E., Prusiner, S.B., Pines, A., and Wemmer, D.E. (2001). Solid-state NMR studies of the secondary structure of a mutant prion protein fragment of 55 residues that induces neurodegeneration. Proc. Natl. Acad. Sci. U.S.A. *98*, 11686–11690.

374) Leclerc, E., Peretz, D., Ball, H., Sakurai, H., Legname, G., Serban, A., Prusiner, S.B., Burton, D.R., and Williamson, R.A. (2001). Immobilized prion protein undergoes spontaneous rearrangement to a conformation having features in common with the infectious form. EMBO J. *20*, 1547–1554.

375) Mastrianni, J.A., Capellari, S., Telling, G.C., Han, D., Bosque, P., Prusiner, S.B., and DeArmond, S.J. (2001). Inherited prion disease caused by the V210I mutation. Neurology *57*, 2198–2205.

376) Matsunaga, Y., Peretz, D., Williamson, A., Burton, D., Mehlhorn, I., Groth, D., Cohen, F.E., Prusiner, S.B., and Baldwin, M.A. (2001). Cryptic epitopes in N-terminally truncated prion protein are exposed in the full-length molecule: Dependence of conformation on pH. Proteins *44*, 110–118.

377) Mo, H., Moore, R.C., Cohen, F.E., Westaway, D., Prusiner, S.B., Wright, P.E., and Dyson, H.J. (2001). Two different neurodegenerative diseases caused by proteins with similar structures. Proc. Natl. Acad. Sci. U.S.A. *98*, 2352–2357.

378) Moore, R.C., Mastrangelo, P., Bouzamondo, E., Heinrich, C., Legname, G., Prusiner, S.B., Hood, L., Westaway, D., DeArmond, S.J., and Tremblay, P. (2001a). Doppel-induced cerebellar degeneration in transgenic mice. Proc. Natl. Acad. Sci. U.S.A. *98*, 15288–15293.

379) Moore, R.C., Xiang, F., Monaghan, J., Han, D., Zhang, Z., Edström, L., Anvret, M., and Prusiner, S.B. (2001b). Huntington disease phenocopy is a familial prion disease. Am. J. Hum. Genet. *69*, 1385–1388.

380) Peretz, D., Scott, M., Groth, D., Williamson, A., Burton, D., Cohen, F.E., and Prusiner, S.B. (2001a). Strain-specified relative conformational stability of the scrapie prion protein. Protein Sci. *10*, 854–863.

381) Peretz, D., Williamson, R. A., Kaneko, K., Vergara, J., Leclerc, E., Schmitt-Ulms, G., Mehlhorn, I. R., Legname, G., Wormald, M. R., Rudd, P. M., Dwek, R. A., Burton, D. R., and Prusiner, S. B. (2001b). Antibodies inhibit prion propagation and clear cell cultures of prion infectivity. Nature *412*, 739–743.

382) Prusiner, S.B. (2001). Shattuck Lecture — Neurodegenerative diseases and prions. N. Engl. J. Med. *344*, 1516–1526.

383) Requena, J.R., Groth, D., Legname, G., Stadtman, E.R., Prusiner, S.B., and Levine, R.L. (2001). Copper-catalyzed oxidation of the recombinant SHa(29–231) prion protein. Proc. Natl. Acad. Sci. U.S.A. *98*, 7170–7175.

384) Rudd, P.M., Wormald, M.R., Wing, D.R., Prusiner, S.B., and Dwek, R.A. (2001). Prion glycoprotein: structure, dynamics, and roles for the sugars. Biochemistry *40*, 3759–3766.

385) Schlumpberger, M., Prusiner, S.B., and Herskowitz, I. (2001). Induction of distinct [*URE3*] yeast prion strains. Mol. Cell. Biol. *21*, 7035–7046.

386) Schmitt-Ulms, G., Legname, G., Baldwin, M.A., Ball, H.L., Bradon, N., Bosque, P.J., Crossin, K.L., Edelman, G.M., DeArmond, S.J., Cohen, F.E., and Prusiner, S.B. (2001). Binding of neural cell adhesion molecules (N-CAMs) to the cellular prion protein. J. Mol. Biol. *314*, 1209–1225.

387) Supattapone, S., Bouzamondo, E., Ball, H.L., Wille, H., Nguyen, H.-O.B., Cohen, F.E., DeArmond, S.J., Prusiner, S.B., and Scott, M. (2001a). A protease-resistant 61-residue prion peptide causes neurodegeneration in transgenic mice. Mol. Cell. Biol. *21*, 2608–2616.

388) Supattapone, S., Muramoto, T., Legname, G., Mehlhorn, I., Cohen, F.E., DeArmond, S.J., Prusiner, S.B., and Scott, M.R. (2001b). Identification of two prion protein regions that modify scrapie incubation time. J. Virol. *75*, 1408–1413.

389) Supattapone, S., Wille, H., Uyechi, L., Safar, J., Tremblay, P., Szoka, F.C., Cohen, F.E., Prusiner, S.B., and Scott, M.R. (2001c). Branched polyamines cure prion-infected neuroblastoma cells. J. Virol. *75*, 3453–3461.

390) Viles, J.H., Donne, D., Kroon, G., Prusiner, S.B., Cohen, F.E., Dyson, H.J., and Wright, P.E. (2001). Local structural plasticity of the prion protein. Analysis of NMR relaxation dynamics. Biochemistry *40*, 2743–2753.

391) Baskakov, I.V., Legname, G., Baldwin, M.A., Prusiner, S.B., and Cohen, F.E. (2002). Pathway complexity of prion protein assembly into amyloid. J. Biol. Chem. *277*, 21140–21148.

392) Bosque, P.J., Ryou, C., Telling, G., Peretz, D., Legname, G., DeArmond, S.J., and Prusiner, S.B. (2002). Prions in skeletal muscle. Proc. Natl. Acad. Sci. U.S.A. *99*, 3812–3817.

393) Kuwata, K., Li, H., Yamada, H., Legname, G., Prusiner, S.B., Akasaka, K., and James, T.L. (2002). Locally disordered conformer of the hamster prion protein: a crucial intermediate to PrP^{Sc}? Biochemistry *41*, 12277–12283.

394) Legname, G., Nelken, P., Guan, Z., Kanyo, Z.F., DeArmond, S.J., and Prusiner, S.B. (2002). Prion and doppel proteins bind to granule cells of the cerebellum. Proc. Natl. Acad. Sci. U.S.A. *99*, 16285–16290.

395) Nicholson, E.M., Mo, H., Prusiner, S.B., Cohen, F.E., and Marqusee, S. (2002). Differences between the prion protein and its homolog doppel: a partially structured state with implications for scrapie formation. J. Mol. Biol. *316*, 807–815.

396) Peretz, D., Williamson, R.A., Legname, G., Matsunaga, Y., Vergara, J., Burton, D., DeArmond, S.J., Prusiner, S.B., and Scott, M.R. (2002). A change in the conformation of prions accompanies the emergence of a new prion strain. Neuron *34*, 921–932.

397) Perrier, V., Kaneko, K., Safar, J., Vergara, J., Tremblay, P., DeArmond, S.J., Cohen, F.E., Prusiner, S.B., and Wallace, A.C. (2002). Dominant-negative inhibition of prion replication in transgenic mice. Proc. Natl. Acad. Sci. U.S.A. *99*, 13079–13084.

398) Prusiner, S.B. (2002). Historical essay. Discovering the cause of AIDS. Science *298*, 1726.

399) Safar, J. G., Scott, M., Monaghan, J., Deering, C., Didorenko, S., Vergara, J., Ball, H., Legname, G., Leclerc, E., Solforosi, L., Serban, H., Groth, D., Burton, D. R., Prusiner, S. B., and Williamson, R. A. (2002). Measuring prions causing bovine spongiform encephalopathy or chronic wasting disease by immunoassays and transgenic mice. Nat. Biotechnol. *20*, 1147–1150.

400) Wille, H., Michelitsch, M.D., Guénebaut, V., Supattapone, S., Serban, A., Cohen, F.E., Agard, D.A., and Prusiner, S.B. (2002). Structural studies of the scrapie prion protein by electron crystallography. Proc. Natl. Acad. Sci. U.S.A. *99*, 3563–3568.

401) Yehiely, F., Bamborough, P., Costa, M.D., Perry, B.J., Thinakaran, G., Cohen, F.E., Carlson, G.A., and Prusiner, S.B. (2002). Identification of candidate proteins binding to prion protein [erratum]. Neurobiol. Dis. *10*, 67–68.

402) Burns, C.S., Aronoff-Spencer, E., Legname, G., Prusiner, S.B., Antholine, W.E., Gerfen, G.J., Peisach, J., and Millhauser, G.L. (2003). Copper coordination in the full-length, recombinant prion protein. Biochemistry *42*, 6794–6803.

403) DeArmond, S.J., and Prusiner, S.B. (2003). Perspectives on prion biology, prion disease pathogenesis, and pharmacologic approaches to treatment. Clin. Lab. Med. *23*, 1–41.

404) Korth, C., Kaneko, K., Groth, D., Heye, N., Telling, G., Mastrianni, J., Parchi, P., Gambetti, P., Will, R., Ironside, J., Heinrich, C., Tremblay, P., DeArmond, S. J., and Prusiner, S. B. (2003). Abbreviated incubation times for human prions in mice expressing a chimeric mouse—human prion protein transgene. Proc. Natl. Acad. Sci. U.S.A. *100*, 4784–4789.

405) Leclerc, E., Peretz, D., Ball, H., Solforosi, L., Legname, G., Safar, J., Serban, A., Prusiner, S.B., Burton, D.R., and Williamson, R.A. (2003). Conformation of PrP^C on the cell surface as probed by antibodies. J. Mol. Biol. *326*, 475–483.

406) May, B.C.H., Fafarman, A.T., Hong, S.B., Rogers, M., Deady, L.W., Prusiner, S.B., and Cohen, F.E. (2003). Potent inhibition of scrapie prion replication in cultured cells by bis-acridines. Proc. Natl. Acad. Sci. U.S.A. *100*, 3416–3421.

407) Mironov, A., Latawiec, D., Wille, H., Bouzamondo-Bernstein, E., Legname, G., Williamson, R.A., Burton, D., DeArmond, S.J., Prusiner, S.B., and Peters, P.J. (2003). Cytosolic prion protein in neurons. J. Neurosci. *23*, 7183–7193.

408) Peters, P.J., Mironov, A., Peretz, D., van Donselaar, E., Leclerc, E., Erpel, S., DeArmond, S.J., Burton, D.R., Williamson, R.A., Vey, M., and Prusiner, S.B. (2003). Trafficking of prion proteins through a caveolae-mediated endosomal pathway. J. Cell Biol. *162*, 703–717.

409) Qin, K., Coomaraswamy, J., Mastrangelo, P., Yang, Y., Lugowski, S., Petromilli, C., Prusiner, S.B., Fraser, P.E., Goldberg, J.M., Chakrabartty, A., and Westaway, D. (2003). The PrP-like protein doppel binds copper. J. Biol. Chem. *278*, 8888–8896.

410) Ryou, C., Legname, G., Peretz, D., Craig, J.C., Baldwin, M.A., and Prusiner, S.B. (2003a). Differential inhibition of prion propagation by enantiomers of quinacrine. Lab. Invest. *83*, 837–843.

411) Ryou, C., Prusiner, S.B., and Legname, G. (2003b). Cooperative binding of dominant-negative prion protein to kringle domains. J. Mol. Biol. *329*, 323–333.

412) Baskakov, I.V., Legname, G., Gryczynski, Z., and Prusiner, S.B. (2004). The peculiar nature of unfolding of the human prion protein. Protein Sci. *13*, 586–595.

413) Bouzamondo-Bernstein, E., Hopkins, S.D., Spilman, P., Uyehara-Lock, J., Deering, C., Safar, J., Prusiner, S.B., Ralston, H.J., III, and DeArmond, S.J. (2004). The neurodegeneration sequence in prion diseases: Evidence from functional, morphological and ultrastructural studies of the GABAergic system. J. Neuropathol. Exp. Neurol. *63*, 882–899.

414) Goldman, J.S., Miller, B.L., Safar, J., de Tourreil, S., Martindale, J.L., Prusiner, S.B., and Geschwind, M.D. (2004). When sporadic disease is not sporadic: the potential for genetic etiology. Arch. Neurol. *61*, 213–216.

415) Govaerts, C., Wille, H., Prusiner, S.B., and Cohen, F.E. (2004). Evidence for assembly of prions with left-handed b-helices into trimers. Proc. Natl. Acad. Sci. U.S.A. *101*, 8342–8347.

416) Legname, G., Baskakov, I.V., Nguyen, H.-O.B., Riesner, D., Cohen, F.E., DeArmond, S.J., and Prusiner, S.B. (2004). Synthetic mammalian prions. Science *305*, 673–676.

417) May, B.C.H., Govaerts, C., Prusiner, S.B., and Cohen, F.E. (2004). Prions: so many fibers, so little infectivity. Trends Biochem. Sci. *29*, 162–165.

418) Prusiner, S.B. (2004a). Detecting mad cow disease. Sci. Am. *291*, 86–93.

419) Prusiner, S.B. (2004b). Early evidence that a protease-resistant protein is an active component of the infectious prion. Cell *S116*, S109.

420) Requena, J.R., Dimitrova, M.N., Legname, G., Teijeira, S., Prusiner, S.B., and Levine, R.L. (2004). Oxidation of methionine residues in the prion protein by hydrogen peroxide. Arch. Biochem. Biophys. *432*, 188–195.

421) Schmitt-Ulms, G., Hansen, K., Liu, J., Cowdrey, C., Yang, J., DeArmond, S.J., Cohen, F.E., Prusiner, S.B., and Baldwin, M.A. (2004). Time-controlled transcardiac perfusion cross-linking for the study of protein interactions in complex tissues. Nat. Biotechnol. *22*, 724–731.

422) Serban, A., Legname, G., Hansen, K., Kovaleva, N., and Prusiner, S.B. (2004). Immunoglobulins in urine of hamsters with scrapie. J. Biol. Chem. *279*, 48817–48820.

423) Tremblay, P., Ball, H.L., Kaneko, K., Groth, D., Hegde, R.S., Cohen, F.E., DeArmond, S.J., Prusiner, S.B., and Safar, J.G. (2004). Mutant PrP^{Sc} conformers induced by a synthetic peptide and several prion strains. J. Virol. *78*, 2088–2099.

424) Yung, L., Huang, Y., Lessard, P., Legname, G., Lin, E.T., Baldwin, M., Prusiner, S.B., Ryou, C., and Guglielmo, B.J. (2004). Pharmacokinetics of quinacrine in the treatment of prion disease. BMC Infect. Dis. *4*, 53–59.

425) Ishikura, N., Clever, J.L., Bouzamondo-Bernstein, E., Samayoa, E., Prusiner, S.B., Huang, E.J., and DeArmond, S.J. (2005). Notch-1 activation and dendritic atrophy in prion disease. Proc. Natl. Acad. Sci. U.S.A. *102*, 886–891.

426) Kanaani, J., Prusiner, S.B., Diacovo, J., Baekkeskov, S., and Legname, G. (2005). Recombinant prion protein induces rapid polarization and development of synapses in embryonic rat hippocampal neurons *in vitro*. J. Neurochem. *95*, 1373–1386.

427) Lee, I.S., Long, J.R., Prusiner, S.B., and Safar, J.G. (2005). Selective precipitation of prions by polyoxometalate complexes. J. Am. Chem. Soc. *127*, 13802–13803.

428) Leffers, K.W., Wille, H., Stohr, J., Junger, E., Prusiner, S.B., and Riesner, D. (2005). Assembly of natural and recombinant prion protein into fibrils. Biol. Chem. *386*, 569–580.

429) Legname, G., Nguyen, H.-O.B., Baskakov, I.V., Cohen, F.E., DeArmond, S.J., and Prusiner, S.B. (2005). Strain-specified characteristics of mouse synthetic prions. Proc. Natl. Acad. Sci. U.S.A. *102*, 2168–2173.

430) Prusiner, S.B. (2005). Remembering Radoslav Andjus. Ann. N.Y. Acad. Sci. *1048*.

431) Safar, J.G., DeArmond, S.J., Kociuba, K., Deering, C., Didorenko, S., Bouzamondo-Bernstein, E., Prusiner, S.B., and Tremblay, P. (2005a). Prion clearance in bigenic mice. J. Gen. Virol. *86*, 2913–2923.

432) Safar, J. G., Geschwind, M. D., Deering, C., Didorenko, S., Sattavat, M., Sanchez, H., Serban, A., Vey, M., Baron, H., Giles, K., Miller, B. L., DeArmond, S. J., and Prusiner, S. B. (2005b). Diagnosis of human prion disease. Proc. Natl. Acad. Sci. U.S.A. *102*, 3501–3506.

433) Safar, J.G., Kellings, K., Serban, A., Groth, D., Cleaver, J.E., Prusiner, S.B., and Riesner, D. (2005c). Search for a prion-specific nucleic acid. J. Virol. *79*, 10796–10806.

434) Scott, M.R., Peretz, D., Nguyen, H.-O.B., DeArmond, S.J., and Prusiner, S.B. (2005). Transmission barriers for bovine, ovine, and human prions in transgenic mice. J. Virol. *79*, 5259–5271.

435) Boy, J., Leergaard, T. B., Schmidt, T., Odeh, F., Bichelmeier, U., Nuber, S., Holzmann, C., Wree, A., Prusiner, S. B., Bujard, H., Riess, O., and Bjaalie, J. G. (2006). Expression mapping of tetracycline-responsive prion protein promoter: Digital atlasing for generating cell-specific disease models. Neuroimage *33*, 449–462.

436) Giri, R.K., Young, R.Y., Pitstick, R., DeArmond, S.J., Prusiner, S.B., and Carlson, G.A. (2006). Prion infection of mouse neurospheres. Proc. Natl. Acad. Sci. U.S.A. *103*, 3875–3880.

437) Huang, Y., Okochi, H., May, B.C., Legname, G., Prusiner, S.B., Benet, L.Z., Guglielmo, B.J., and Lin, E.T. (2006). Quinacrine is mainly metabolized to mono-desethyl quinacrine by CYP3A4/5 and its brain accumulation is limited by P-glycoprotein. Drug Metab. Dispos. *34*, 1136–1144.

438) Leclerc, E., Serban, H., Prusiner, S.B., Burton, D.R., and Williamson, R.A. (2006). Copper induces conformational changes in the N-terminal part of cell-surface PrP(C). Arch. Virol. *151*, 2103–2109.

439) Legname, G., Nguyen, H.-O.B., Peretz, D., Cohen, F.E., DeArmond, S.J., and Prusiner, S.B. (2006). Continuum of prion protein structures enciphers a multitude of prion isolate-specified phenotypes. Proc. Natl. Acad. Sci. U.S.A. *103*, 19105–19110.

440) Lim, K.H., Nguyen, T.N., Damo, S.M., Mazur, T., Ball, H.L., Prusiner, S.B., Pines, A., and Wemmer, D.E. (2006). Solid-state NMR structural studies of the fibril form of a mutant mouse prion peptide PrP(89–143)(P101L). Solid State Nucl. Magn. Reson. *29*, 183–190.

441) Luginbuhl, B., Kanyo, Z., Jones, R.M., Fletterick, R.J., Prusiner, S.B., Cohen, F.E., Williamson, R.A., Burton, D.R., and Pluckthun, A. (2006). Directed evolution of an anti-prion protein scFv fragment to an affinity of 1 pM and its structural interpretation. J. Mol. Biol. *363*, 75–97.

442) May, B.C.H., Witkop, J., Sherrill, J., Anderson, M.O., Madrid, P.B., Zorn, J.A., Prusiner, S.B., Cohen, F.E., and Guy, R.K. (2006). Structure-activity relationship study of 9-aminoacridine compounds in scrapie-infected neuroblastoma cells. Bioorg. Med. Chem. Lett. *16*, 4913–4916.

443) Peretz, D., Supattapone, S., Giles, K., Vergara, J., Freyman, Y., Lessard, P., Safar, J. G., Glidden, D. V., McCulloch, C., Nguyen, H.-O. B., Scott, M., DeArmond, S. J., and Prusiner, S. B. (2006). Inactivation of prions by acidic sodium dodecyl sulfate. J. Virol. *80*, 322–331.

444) Prusiner, S.B., and McCarty, M. (2006). Discovering DNA encodes heredity and prions are infectious proteins. Annu. Rev. Genet. *40*, 25–45.

445) Safar, J. G., Wille, H., Geschwind, M. D., Deering, C., Latawiec, D., Serban, A., King, D. J., Legname, G., Weisgraber, K. H., Mahley, R. W., Miller, B. L., DeArmond, S. J., and Prusiner, S. B. (2006). Human prions and plasma lipoproteins. Proc. Natl. Acad. Sci. U.S.A. *103*, 11312–11317.

446) Tamgüney, G., Giles, K., Bouzamondo-Bernstein, E., Bosque, P.J., Miller, M.W., Safar, J., DeArmond, S.J., and Prusiner, S.B. (2006). Transmission of elk and deer prions to transgenic mice. J. Virol. *80*, 9104–9114.

447) Colby, D.W., Zhang, Q., Wang, S., Groth, D., Legname, G., Riesner, D., and Prusiner, S.B. (2007). Prion detection by an amyloid seeding assay. Proc. Natl. Acad. Sci. U.S.A. *104*, 20914–20919.

448) Diez, M., Groth, D., DeArmond, S.J., Prusiner, S.B., and Hokfelt, T. (2007). Changes in neuropeptide expression in mice infected with prions. Neurobiol. Aging *28*, 748–765.

449) Ghaemmaghami, S., Phuan, P.W., Perkins, B., Ullman, J., May, B.C., Cohen, F.E., and Prusiner, S.B. (2007). Cell division modulates prion accumulation in cultured cells. Proc. Natl. Acad. Sci. U.S.A. *104*, 17971–17976.

450) Karpuj, M.V., Giles, K., Gelibter-Niv, S., Scott, M.R., Lingappa, V.R., Szoka, F.C., Peretz, D., Denetclaw, W., and Prusiner, S.B. (2007). Phosphorothioate oligonucleotides reduce PrP^{Sc} levels and prion infectivity in cultured cells. Mol. Med. *13*, 190–198.

451) King, D.J., Safar, J.G., Legname, G., and Prusiner, S.B. (2007). Thioaptamer interactions with prion proteins: sequence-specific and non-specific binding sites. J. Mol. Biol. *369*, 1001–1014.

452) May, B.C.H., Zorn, J.A., Witkop, J., Sherrill, J., Wallace, A.C., Legname, G., Prusiner, S.B., and Cohen, F.E. (2007). Structure-activity relationship study of prion inhibition by 2-aminopyridine-3,5-dicarbonitrile-based compounds: Parallel synthesis, bioactivity and in vitro pharmacokinetics. J. Med. Chem. *50*, 65–73.

453) Muller, H., Stitz, L., Wille, H., Prusiner, S.B., and Riesner, D. (2007). Influence of water, fat, and glycerol on the mechanism of thermal prion inactivation. J. Biol. Chem. *282*, 35855–35867.

454) Norrby, E., and Prusiner, S.B. (2007). Polio and Nobel Prizes: looking back 50 years. Ann. Neurol. *61*, 385–395.

455) Philipp, W. J., Groth, D., Giles, K., Vodrazka, P., Schimmel, H., Feyssaguet, M., Toomik, R., Schacher, P., Osman, A., Lachmann, I., Wear, A., Arsac, J.-N., and Prusiner, S. B. (2007). Transgenic mouse brains for evaluation and quality control of BSE tests. Biol. Chem. *388*, 349–354.

456) Phuan, P.-W., Zorn, J.A., Safar, J., Giles, K., Prusiner, S.B., Cohen, F.E., and May, B.C.H. (2007). Discriminating between cellular and misfolded prion protein by using affinity to 9-aminoacridine compounds. J. Gen. Virol. *88*, 1392–1401.

457) Tremblay, P., Bouzamondo-Bernstein, E., Heinrich, C., Prusiner, S.B., and DeArmond, S.J. (2007). Developmental expression of PrP in the post-implantation embryo. Brain Res. *1139*, 60–67.

458) Wille, H., Govaerts, C., Borovinskiy, A., Latawiec, D., Downing, K.H., Cohen, F.E., and Prusiner, S.B. (2007). Electron crystallography of the scrapie prion protein complexed with heavy metals. Arch. Biochem. Biophys. *467*, 239–248.

459) Feng, B.Y., Toyama, B.H., Wille, H., Colby, D.W., Collins, S.R., May, B.C.H., Prusiner, S.B., Weissman, J., and Shoichet, B.K. (2008). Small-molecule aggregates inhibit amyloid polymerization. Nat. Chem. Biol. *4*, 197–199.

460) Giles, K., Glidden, D.V., Beckwith, R., Seoanes, R., Peretz, D., DeArmond, S.J., and Prusiner, S.B. (2008). Resistance of bovine spongiform encephalopathy (BSE) prions to inactivation. PLoS Pathog. *4*, e1000206, 1–9.

461) Godsave, S.F., Wille, H., Kujala, P., Latawiec, D., DeArmond, S.J., Serban, A., Prusiner, S.B., and Peters, P.J. (2008). Cryo-immunogold electron microscopy for prions: toward identification of a conversion site. J. Neurosci. *28*, 12489–12499.

462) Prusiner, S.B. (2008). Reflections on kuru. Philos. Trans. R. Soc. B *363*, 3654–3656.

463) Safar, J.G., Lessard, P., Tamgüney, G., Freyman, Y., Deering, C., Letessier, F., DeArmond, S.J., and Prusiner, S.B. (2008). Transmission and detection of prions in feces. J. Infect. Dis. *198*, 81–89.

464) Spilman, P., Lessard, P., Sattavat, M., Bush, C., Tousseyn, T., Huang, E. J., Giles, K., Golde, T., Das, P., Fauq, A., Prusiner, S. B., and DeArmond, S. J. (2008). A gamma-secretase inhibitor and quinacrine reduce prions and prevent dendritic degeneration in murine brains. Proc. Natl. Acad. Sci. U.S.A. *105*, 10595–10600.

465) Stöhr, J., Weinmann, N., Wille, H., Kaimann, T., Nagel-Steger, L., Birkmann, E., Panza, G., Prusiner, S.B., Eigen, M., and Riesner, D. (2008). Mechanisms of prion protein assembly into amyloid. Proc. Natl. Acad. Sci. U.S.A. *105*, 2409–2414.

466) Tamgüney, G., Giles, K., Glidden, D. V., Lessard, P., Wille, H., Tremblay, P., Groth, D. F., Yehiely, F., Korth, C., Moore, R. C., Tatzelt, J., Rubinstein, E., Boucheix, C., Yang, X., Stanley, P., Lisanti, M. P., Dwek, R. A., Rudd, P. M., Moskovitz, J., Epstein, C. J., Dawson Cruz, T., Kuziel, W. A., Maeda, N., Sap, J., Hsiao Ashe, K., Carlson, G. A., Tesseur, I., Wyss-Coray, T., Mucke, L., Weisgraber, K. H., Mahley, R. W., Cohen, F. E., and Prusiner, S. B. (2008). Genes contributing to prion pathogenesis. J. Gen. Virol. *89*, 1777–1788.

467) Bae, S.-H., Legname, G., Serban, A., Prusiner, S.B., Wright, P.E., and Dyson, H.J. (2009). Prion proteins with pathogenic and protective mutations show similar structure and dynamics. Biochemistry *48*, 8120–8128.

468) Campana, V., Zentilin, L., Mirabile, I., Kranjc, A., Casanova, P., Giacca, M., Prusiner, S.B., Legname, G., and Zurzolo, C. (2009). Development of antibody fragments for immunotherapy of prion diseases. Biochem. J. *418*, 507–515.

469) Choi, E.M., Geschwind, M.D., Deering, C., Pomeroy, K., Kuo, A., Miller, B.L., Safar, J.G., and Prusiner, S.B. (2009). Prion proteins in subpopulations of white blood cells from patients with sporadic Creutzfeldt-Jakob disease. Lab. Invest. *89*, 624–635.

470) Colby, D.W., Giles, K., Legname, G., Wille, H., Baskakov, I.V., DeArmond, S.J., and Prusiner, S.B. (2009). Design and construction of diverse mammalian prion strains. Proc. Natl. Acad. Sci. U.S.A. *106*, 20417–20422.

471) Ghaemmaghami, S., Ahn, M., Lessard, P., Giles, K., Legname, G., DeArmond, S.J., and Prusiner, S.B. (2009). Continuous quinacrine treatment results in the formation of drug-resistant prions. PLoS Pathog. *5*, e1000673, 1–10.

472) Olanow, C.W., and Prusiner, S.B. (2009). Is Parkinson's disease a prion disorder? Proc. Natl. Acad. Sci. U.S.A. *106*, 12571–12572.

473) Tamgüney, G., Francis, K.P., Giles, K., Lemus, A., DeArmond, S.J., and Prusiner, S.B. (2009a). Measuring prions by bioluminescence imaging. Proc. Natl. Acad. Sci. U.S.A. *106*, 15002–15006.

474) Tamgüney, G., Miller, M.W., Giles, K., Lemus, A., Glidden, D.V., DeArmond, S.J., and Prusiner, S.B. (2009b). Transmission of scrapie and sheep-passaged bovine spongiform encephalopathy prions to transgenic mice expressing elk prion protein. J. Gen. Virol. *90*, 1035–1047.

475) Tamgüney, G., Miller, M.W., Wolfe, L.L., Sirochman, T.M., Glidden, D.V., Palmer, C., Lemus, A., DeArmond, S.J., and Prusiner, S.B. (2009c). Asymptomatic deer excrete infectious prions in faeces. Nature *461*, 529–532.

476) Wille, H., Bian, W., McDonald, M., Kendall, A., Colby, D.W., Bloch, L., Ollesch, J., Boronvinskiy, A.L., Cohen, F.E., Prusiner, S.B., and Stubbs, G. (2009a). Natural and synthetic prion structure from X-ray fiber diffraction. Proc. Natl. Acad. Sci. U.S.A. *106*, 16990–16995.

477) Wille, H., Shanmugam, M., Murugesu, M., Ollesch, J., Stubbs, G., Long, J.R., Safar, J.G., and Prusiner, S.B. (2009b). Surface charge of polyoxometalates modulates polymerization of the scrapie prion protein. Proc. Natl. Acad. Sci. U.S.A. *106*, 3740–3745.

478) Colby, D.W., Wain, R., Baskakov, I.V., Legname, G., Palmer, C.G., Nguyen, H.-O.B., Lemus, A., Cohen, F.E., DeArmond, S.J., and Prusiner, S.B. (2010). Protease-sensitive synthetic prions. PLoS Pathog. *6*, e1000736.

479) Doak, A.K., Wille, H., Prusiner, S.B., Shoichet, B.K. (2010). Colloid formation by drugs in simulated intestinal fluid. J. Med. Chem. *53*, 4259–4265.

480) Ghaemmaghami, S., May, B.C.H., Renslo, A.R., and Prusiner, S.B. (2010). Discovery of 2-aminothiazoles as potent antiprion compounds. J. Virol. *84*, 3408–3412.

481) Ghaemmaghami, S., Ullman, J., Ahn, M., St. Martin, S., and Prusiner, S.B. (2010). Chemical induction of misfolded prion protein conformers in cell culture. J. Biol. Chem., *285*, 10415–10423.

482) Giles, K., Glidden, D.V., Patel, S., Korth, C., Groth, D., Lemus, A., DeArmond, S.J., Prusiner, S.B. (2010) Human prion strain selection in transgenic mice. Ann. Neurol. *68*, 151–161.

483) Hnasko, R., Serban, A.V., Carlson, G.A., Prusiner, S.B., Stanker, L.H. (2010). Generation of antisera to purified prions in lipid rafts. Prion. *4,* 94–104.

484) Price, J.C., Guan, S., Burlingame, A.L., Prusiner, S.B., Ghaemmaghami, S., (2010). Analysis of proteome dynamics in the mouse brain. Proc. Natl. Acad. Sci. U.S.A.. *107*, 14508–14513.

485) Stanker, L.H., Serban, A.V., Cleveland, E., Hnasko, R., Lemus, A., Safer, J.G., DeArmond, S.J., Prusiner, S.B. (2010). Conformation-dependent high-affinity mAbs to prion proteins. J. Immunol. *185,* 729–737.

486) Tamgüney, G., Miller, M.W., Wolfe, L.L., Sirochman, T.M., Glidden, D.V., Palmer, C., Lemus, A., DeArmond, S.J., and Prusiner, S.B. (2010). Corrigendum: Asymptomatic deer excrete infectious prions in faeces. Nature. *466*, 652.

487) Gallardo-Godoy, A., Gever, J., Fife, K.L., Silber, B.M., Prusiner, S.B., and Renslo, A.R. (2011). 2-Aminothiazoles as therapeutic leads for prion diseases. J. Med. Chem. *54*, 1010–1021.

488) Ghaemmaghami, S., Watts, J. C., Nguyen, H.-O., Hayashi, S., DeArmond, S.J., and Prusiner, S.B. (2011). Conformational transformation and selection of synthetic prion strains. J. Mol. Biol. *413*, 527–542.

489) Guan, S., Price, J.C., Prusiner, S.B., Ghaemmaghami, S., and Burlingame, A.L. (2011). A data processing pipeline for mammalian proteome dynamics studies using stable isotope metabolic labeling. Mol. Cell. Proteomics. *10*, M111.010728.

490) Hnasko, R., Serban, A.V., Carlson, G., Prusiner, S.B., and Stanker, L.H. (2011). Generation of antisera to purified prions in lipid rafts [erratum]. Prion. *5*, 235.

491) Kujala, P., Raymond, C. R., Romeijn, M., Godsave, S.F., van Kasteren, S.I., Wille, H., Prusiner, S.B., Mabbott, N.A., and Peters, P.J. (2011). Prion uptake in the gut: identification of the first uptake and replication sites. PLoS. Pathog. *7*, e1002449, 1–19.

492) Poncet-Montange, G., St.-Martin, S.J., Bogatova, O.V., Prusiner, S.B., Shoichet, B.K., and Ghaemmaghami, S. (2011). A survey of antiprion compounds reveals the prevalence of non-PrP molecular targets. J. Biol. Chem. *286*, 27718–27728.

493) Safar, J.G., Giles, K., Lessard, P., Letessier, F., Patel, S., Serban, A., DeArmond, S.J., and Prusiner, S.B. (2011). Conserved properties of human and bovine prion strains on transmission to guinea pigs. Lab. Invest. *91*, 1326–1336.

494) Stöhr, J., Watts, J.C., Legname, G., Oehler, A., Lemus, A., Nguyen, H.-O. B., Sussman, J., Wille, H., DeArmond, S.J., Prusiner, S.B., and Giles, K. (2011). Spontaneous generation of anchorless prions in transgenic mice. Proc. Natl. Acad. Sci. U.S.A. *108*, 21223–21228.

495) Watts, J.C., Giles, K., Grillo, S.K., Lemus, A., DeArmond, S.J., and Prusiner, S.B. (2011a). Bioluminescence imaging of Abeta deposition in bigenic mouse models of Alzheimer's disease. Proc. Natl. Acad. Sci. U.S.A. *108*, 2528–2533.

496) Watts, J.C., Stöhr, J., Bhardwaj, S., Wille, H., Oehler, A., DeArmond, S.J., Giles, K., and Prusiner, S.B. (2011b). Protease-resistant prions selectively decrease shadoo protein. PLoS. Pathog. *7*, e1002382, 1–16.

497) Ahn, M., Ghaemmaghami, S., Huang, Y., Phuan, P.-W., May, B.C.H., Giles, K., DeArmond, S.J., and Prusiner, S.B. (2012). Pharmacokinetics of quinacrine efflux from mouse brain via the P-glycoprotein efflux transporter. PLoS. ONE *7*, e39112, 1–7.

498) Giles, K., De Nicola, G.F., Patel, S., Glidden, D.V., Korth, C., Oehler, A., DeArmond, S.J., and Prusiner, S.B. (2012). Identifying I137M and other mutations that modulate incubation periods for two human prion strains. J. Virol. *86*, 6033–6041.

499) Guan, S., Price, J.C., Ghaemmaghami, S., Prusiner, S.B., and Burlingame, A.L. (2012). Compartment modeling for mammalian protein turnover studies by stable isotope metabolic labeling. Anal. Chem. *84*, 4014–4021.

500) Nazor Friberg, K., Hung, G., Wancewicz, E., Giles, K., Black, C., Freier, S., Bennett, F., DeArmond, S.J., Freyman, Y., Lessard, P., Ghaemmaghami, S., and Prusiner, S.B. (2012). Intracerebral infusion of antisense oligonucleotides into prion-infected mice. Mol. Ther. Nucleic Acids *1*, e9, 1–12.

501) Prusiner, S.B. (2012). A unifying role for prions in neurodegenerative diseases. Science. *1*, 1511–1513.

502) Stanker, L.H., Scotcher, M.C., Lin, A., McGarvey, J., Prusiner, S.B., and Hnasko, R. (2012). Novel epitopes identified by anti-PrP monoclonal antibodies produced following immunization of *Prnp*$^{0/0}$ Balb/cJ mice with purified scrapie prions. Hybridoma. *31*, 314–324.

503) Stöhr, J., Watts, J.C., Mensinger, Z.L., Oehler, A., Grillo, S.K., DeArmond, S.J., Prusiner, S.B., and Giles, K. (2012). Purified and synthetic Alzheimer's amyloid beta (Aβ) prions. Proc. Natl. Acad. Sci. U.S.A. *109*, 11025–11030.

504) Tamgüney, G., Richt, J.A., Hamir, A.N., Greenlee, J.J., Miller, M.W., Wolfe, L.L., Sirochman, T.M., Young, A.J., Glidden, D.V., Johnson, N.L., Giles, K., DeArmond, S.J., and Prusiner, S.B. (2012). Salivary prions in sheep and deer. Prion. *6*, 52–61.

505) Wan, W., Wille, H., Stöhr, J., Baxa, U., Prusiner, S.B., and Stubbs, G. (2012). Degradation of fungal prion HET-s(218–289) induces formation of a generic amyloid fold. Biophys. J. *102*, 2339–2344.

506) Watts, J.C., Giles, K., Stöhr, J., Oehler, A., Bhardwaj, S., Grillo, S.K., Patel, S., DeArmond, S.J., and Prusiner, S.B. (2012). Spontaneous generation of rapidly transmissible prions in transgenic mice expressing wild-type bank vole prion protein. Proc. Natl. Acad. Sci. U.S.A. *109*, 3498–3503.

507) Tamgüney, G., Giles, K., Oehler, A., Johnson, N.L., DeArmond, S.J., and Prusiner, S.B. (2013). Chimeric elk/mouse prion proteins in transgenic mice. J. Gen. Virol. *94*, 443–452.

508) Ghaemmaghami, S., Colby, D.W., Nguyen, H.-O.B., Hayashi, S., Oehler, A., DeArmond, S.J., and Prusiner, S B. (2013). Convergent replication of mouse synthetic prion strains. Am. J. Pathol. *182*, 866–874.

509) Silber, B.M., Rao, S., Fife, K.L., Gallardo-Godoy, A., Renslo, A.R., Dalvie, D.K., Giles, K., Freyman, Y., Elepano, M., Gever, J.R., Li, Z., Jacobson, M.P., Huang, Y., Benet, L.Z., and Prusiner, S.B. (2013). Pharmacokinetics and metabolism of 2-aminothiazoles with antiprion activity in mice. Pharm. Res. *30*, 932–950.

510) Godsave, S.F., Wille, H., Pierson, J., Prusiner, S.B., and Peters, P.J. (2013). Plasma membrane invaginations containing clusters of full-length PrP^{Sc} are an early form of prion-associated neuropathology in vivo. Neurobiol. Aging *34*, 1621–1631.

511) Li, Z., Silber, B.M., Rao, S., Gever, J.R., Bryant, C., Gallardo-Godoy, A., Dolghih, E., Widjaja, K., Elepano, M., Jacobson, M.P., Prusiner, S.B., and Renslo, A.R. (2013). 2-Aminothiazoles with improved pharmacotherapeutic properties for treatment of prion disease. Chem. Med. Chem. *8*, 847–857.

512) Li, Z., Gever, J.R., Rao, S., Widjaja, K., Prusiner, S.B., and Silber, B.M. (2013). Discovery and preliminary structure-activity relationship of arylpiperazines as novel, brain-penetrant antiprion compounds. ACS Med. Chem. Lett. *4*, 397–401.

513) Li, Z., Rao, S., Gever, J. R., Widjaja, K., Prusiner, S.B., and Silber, B.M. (2013). Towards optimization of arylamides as novel, potent, and brain-penetrant antiprion lead compounds. ACS Med. Chem. Lett. *4*, 647–650.

514) Dehdashti, S.J., Zheng, W., Gever, J.R., Wilhelm, R., Nguyen, D.-T., Sittampalam, G., McKew, J.C., Austin, C.P., and Prusiner, S.B. (2013). A high-throughput screening assay for determining cellular levels of total tau protein. Curr. Alzheimer Res. *10*, 679–687.

515) Berry, D.B., Lu, D., Geva, M., Watts, J.C., Bhardwaj, S., Oehler, A., Renslo, A.R., DeArmond, S.J., Prusiner, S.B., and Giles, K. (2013). Drug resistance confounding prion therapeutics. Proc. Natl. Acad. Sci. U.S.A. *110*, E4160–E4169.

516) Lu, D., Giles, K., Li, Z., Rao, S., Dolghih, E., Gever, J.R., Geva, M., Elepano, M.L., Oehler, A., Bryant, C., Renslo, A.R., Jacobson, M.P., DeArmond, S.J., Silber, B.M., and Prusiner, S.B. (2013). Biaryl amides and hydrazones as therapeutics for prion disease in transgenic mice. J. Pharmacol. Exp. Ther. *347*, 325–338.

517) Silber, B.M., Gever, J.R., Li, Z., Gallardo-Godoy, A., Renslo, A.R., Widjaja, K., Irwin, J.J., Rao, S., Jacobson, M.P., Ghaemmaghami, S., and Prusiner, S.B. (2013). Antiprion compounds that reduce PrP^{Sc} levels in dividing and stationary-phase cells. Bioorg. Med. Chem. *21*, 7999–8012.

518) Watts, J.C., Giles, K., Oehler, A., Middleton, L., Dexter, D.T., Gentleman, S.M., DeArmond, S.J., and Prusiner, S.B. (2013). Transmission of multiple system atrophy prions to transgenic mice. Proc. Natl. Acad. Sci. U.S.A. *110*, 19555–19560.

519) Geschwind, M.D., Kuo, A.L., Wong, K.S., Haman, A., Devereaux, G., Raudabaugh, B.J., Johnson, D.Y., Torres-Chae, C.C., Finley, R., Garcia, P., Thai, J.N., Cheng, H.Q., Neuhaus, J.M., Forner, S.A., Duncan, J.L., Possin, K.L., DeArmond, S.J., Prusiner, S.B., and Miller, B.L. (2013). Quinacrine treatment trial for sporadic Creutzfeldt-Jakob disease. Neurology *81*, 2015–2023.

520) Silber, B.M., Gever, J.R., Rao, S., Li, Z., Renslo, A.R., Widjaja, W., Wong, C., Giles, K., Freyman, Y., Elepano, M., Irwin, J.J., Jacobson, M.P., and Prusiner, S.B. (2014). Novel compounds lowering the cellular isoform of the human prion protein in cultured human cells Bioorg. Med. Chem. *22*, 1960-1972.

521) Watts, J. C., Giles, K., Patel, S., Oehler, A., DeArmond, S. J., and Prusiner, S. B. (2014). Evidence that bank vole PrP is a universal acceptor for prions. PLoS Pathog. *10*, e1003990, 1–14.

522) Watts, J.C., Condello, C., Stöhr, J., Oehler, A., Lee, J., DeArmond, S.J., Lannfelt, L., Ingelsson, M., Giles, K., and Prusiner, S.B. (2014). Serial propagation of distinct strains of Aβ prions from Alzheimer's disease patients. Proc. Natl. Acad. Sci. U.S.A. *111*, 10323–10328.

523) Stöhr, J., Condello, C., Watts, J.C., Bloch, L., Oehler, A., Nick, M., DeArmond, S.J., Giles, K., DeGrado, W.F., and Prusiner, S.B. (2014). Distinct synthetic Aβ prion strains producing different amyloid deposits in bigenic mice. Proc. Natl. Acad. Sci. U.S.A. *111*, 10329–10334.

524) Watts, J.C., and Prusiner, S.B. (2014) Mouse models for studying the formation and propagation of prions. J. Biol. Chem. *289*, 19841–19849.

525) Wan, W., Wille, H., Stöhr, J., Kendall, A., Bian, W., McDonald, M., Tiggelaar, S., Watts, J.C., Prusiner, S.B., and Stubbs, G. (2015) Structural studies of truncated forms of the prion protein PrP. Biophys. J. *108*, 1548–1554.

526) Berry, D., Giles, K., Oehler, A., Bhardwaj, S., DeArmond, S.J., and Prusiner, S.B. (2015) Use of a 2-aminothiazole to treat chronic wasting disease in transgenic mice. J. Infect. Dis. *212*, S17–S25.

527) Levine, D.J., Stöhr, J., Falese, L.E., Ollesch, J., Wille, H., Prusiner, S.B., and Long, J.R. (2015) Mechanism of scrapie prion precipitation with phosphotungstate anions. ACS Chem. Biol. *10*, 1269–1277.

528) Watts, J.C., Giles, K., Serban, A., Patel, S., Oehler, A., Bhardwaj, S., Guan, Y., Greicius, M., Miller, B.L., DeArmond, S.J., Geschwind, M.D., and Prusiner, S.B. (2015) Modulation of Creutzfeldt-Jakob disease prion propagation by the A224V mutation. Ann. Neurol. *78*, 540–553.

529) Carter, L., Kim, S.J., Schneidman-Duhovny, D., Stöhr, J., Poncet-Montange, G., Weiss, T.M., Tsuruta, H., Prusiner, S.B., and Sali, A. (2015) Prion protein antibody complexes characterized by chromatography-coupled small-angle X-ray scattering. Biophs. J. *109*, 793–805.

530) Giles, K., Berry, D.B., Condello, C., Hawley, R.C., Gallardo-Godoy, A., Bryant, C., Oehler, A., Elepano, M., Bhardwaj, S., Patel, S., Silber, B.M., Guan, S., DeArmond, S.J., Renslo, A.R., and Prusiner, S.B. (2015) Different 2-aminothiazole therapeutics produce distinct patterns of scrapie prion neuropathology in mouse brains. J. Pharmacol. Exp. Ther. *355*, 2–12.

531) Prusiner, S.B., Woerman, A.L., Rampersaud, R., Watts, J.C., Berry, D.B., Patel, S., Oehler, A., Lowe, J.K., Kravitz, S.N., Geschwind, D.H., Glidden, D.V., Halliday, G., Middleton, L.T., Gentleman, S.M., Mordes, D.A., DeArmond, S.J., and Giles, K. (2015) Evidence for α-synuclein prions causing multiple system atrophy in humans with signs of Parkinson's disease. Proc. Natl. Acad. Sci. U.S.A. *112*, E5308–E5317.

532) Woerman, A. L., Stöhr, J., Aoyagi, A., Rampersaud, R., Krejciova, Z., Watts, J.C., Ohyama, T., Patel, S., Widjaja, K., Oehler, A., Sanders, D.W., Diamond, M.I., Seeley, W.W., Middleton, L., Gentleman, S., Mordes, D.A., Südhof, T.C., Giles, K., and Prusiner, S.B. (2015) Propagation of prions causing synucleinopathies in cultured cells. Proc. Natl. Acad. Sci. U.S.A. *112*, E4949–E4958.

533) Ahlenius, H., Chanda, S., Webb, A.E., Yousif, I., Karmazin, J., Prusiner, S.B., Brunet, A., Südhof, T.C., and Wernig, M. (2016) FoxO3 regulates neuronal reprogramming of cells from postnatal and aging mice. Proc. Natl. Acad. Sci. U.S.A. *113*, 8514–8519.

534) Elkins, M.R., Wang, T., Nick, M., Jo, H., Lemmin, T., Prusiner, S.B., DeGrado, W.F., Stöhr, J., and Hong, M. (2016) Structural polymorphism of Alzheimer's β-amyloid fibrils as controlled by an E22 switch: A solid-state NMR study. J. Am. Chem. Soc. *138*, 9840–9852.

535) Giles, K., Berry, D.B., Condello, C., Dugger, B.N., Li, Z., Oehler, A., Bhardwaj, S., Elepano, M., Guan, S., Silber, B.M., Olson, S.H., and Prusiner, S.B. (2016) Optimization of aryl amides that extend survival in prion-infected mice. J. Pharmacol. Exp. Ther. *358*, 537–547.

536) Watts, J.C., Giles, K., Bourkas, M.E.C., Patel, S., Oehler, A., Gavidia, M., Bhardwaj, S., Lee, J., and Prusiner, S.B. (2016) Towards authentic transgenic mouse models of heritable PrP prion diseases. Acta Neuropathol. *132*, 593–610.

537) Watts, J.C., Giles, K., Saltzberg, D.J., Dugger, B.N., Patel, S., Oehler, A., Bhardwaj, S., Sali, A., and Prusiner, S.B. (2016) Guinea pig prion protein supports rapid propagation of bovine spongiform encephalopathy and variant Creutzfeldt-Jakob disease prions. J. Virol. *90*, 9558–9569.

538) Woerman, A.L., Aoyagi, A., Patel, S., Kazmi, S.A., Lobach, I., Grinberg, L.T., McKee, A.C., Seeley, W.W., Olson, S.H., and Prusiner, S.B. (2016) Tau prions from Alzheimer's disease and chronic traumatic encephalopathy patients propagate in cultured cells. Proc. Natl. Acad. Sci. U.S.A. *113*, E8187–E8196.

539) Stöhr, J., Wu, H., Nick, M., Wu, Y., Bhate, M., Condello, C., Johnson, N., Rodgers, J., Lemmin, T., Acharya, S., Becker, J., Robinson, K., Kelly, M.J.S., Gai, F., Stubbs, G., Prusiner, S.B., and DeGrado, W.F. (2017) A 31-residue peptide induces aggregation of tau's microtubule-binding region in cells. Nat. Chem. *9*, 874–881.

540) Giles, K., Olson, S.H., and Prusiner, S.B. (2017) Developing therapeutics for PrP prion diseases. Cold Spring Harb. Perspect. Med. *7*, a023747, 1–19.

541) Giles, K., Woerman, A.L., Berry, D.B., and Prusiner, S.B. (2017) Bioassays and inactivation of prions. Cold Spring Harb. Perspect. Biol. *9*, a023499, 1–16.

542) Watts, J.C., and Prusiner, S.B. (2017) β-amyloid prions and the pathobiology of Alzheimer's disease. Cold Spring Harb. Perspect. Med. *8*, a023507, 1–14.

543) Watts, J.C., and Prusiner, S.B. (2017) Experimental models of inherited PrP prion diseases. Cold Spring Harb. Perspect. Med. *7*, a027151, 1–15.

544) Woerman, A.L., Watts, J.C., Aoyagi, A., Giles, K., Middleton, L.T., and Prusiner, S.B. (2017) α-Synuclein: Multiple system atrophy prions. Cold Spring Harb. Perspect. Med. *8*, a024588, 1–12.

545) Johnson, N.R., Condello, C., Guan, S., Oehler, A., Becker, J., Gavidia, M., Carlson, G.A., Giles, K., and Prusiner, S.B. (2017) Evidence for sortilin modulating regional accumulation of human tau prions in transgenic mice. Proc. Natl. Acad. Sci. U.S.A. *114*, E11029–E11036.

546) Lopez, T.L., Giles, K., Dugger, B.N., Oehler, A., Condello, C., Krejciova, Z., Castaneda, J.A., Carlson, G.A., and Prusiner, S.B. (2017) A novel vector for transgenesis in the rat CNS. Acta Neuropathol. Commun. *5*, 84, 1–14.

547) Stöhr, J., Wu, H., Nick, M., Wu, Y., Bhate, M., Condello, C., Johnson, N., Rodgers, J., Lemmin, T., Acharya, S., Becker, J., Robinson, K., Kelly, M.J.S., Gai, F., Stubbs, G., Prusiner, S.B., and DeGrado, W.F. (2017) A 31-residue peptide induces aggregation of tau's microtubule-binding region in cells. Nat. Chem. *9*, 874–881.

548) Woerman, A.L., Patel, S., Kazmi, S.A., Oehler, A., Freyman, Y., Espiritu, L., Cotter, R., Castaneda, J.A., Olson, S.H., and Prusiner, S.B. (2017) Kinetics of human mutant tau prion formation in the brains of two transgenic mouse lines. JAMA Neurol. *74*, 1464–1472.

549) Condello, C., Lemmin, T., Stöhr, J., Nick, M., Wu, Y., Watts, J.C., Oehler, A., Keene, C.D., Bird, T.D., van Duinen, S.G., Lannfelt, L., Ingelsson, M., Graff, C., Giles, K., DeGrado, W.F., and Prusiner, S.B. (2018) Structural heterogeneity and intersubject variability of Aβ in familial and sporadic Alzheimer's disease. Proc. Natl. Acad. Sci. U.S.A. *115*, E782–E791.

550) Nick, M., Wu, Y., Schmidt, N.W., Prusiner, S.B., Stöhr, J., and DeGrado, W.F. (2018) A long-lived aβ oligomer resistant to fibrillization. Biopolymers *109*, e23096, 1–9.

551) Woerman, A.L., Kazmi, S.A., Patel, S., Aoyagi, A., Oehler, A., Widjaja, K., Mordes, D.A., Olson, S.H., and Prusiner, S.B. (2018) Familial Parkinson's point mutation abolishes multiple system atrophy prion replication. Proc. Natl. Acad. Sci. U.S.A. *115*, 409–414.

552) Woerman, A.L., Kazmi, S.A., Patel, S., Freyman, Y., Oehler, A., Aoyagi, A., Mordes, D.A., Halliday, G.M., Middleton, L.T., Gentleman, S.M., Olson, S.H., and Prusiner, S.B. (2018) MSA prions exhibit remarkable stability and resistance to inactivation. Acta Neuropathol. *135*, 49–63.

553) Aoyagi, A., Condello, C., Stöhr, J., Yue, W., Lee, J.C., Rivera, B.M., Woerman, A.L., Halliday, G., van Duinen, S., Ingelsson, M., Lannfelt, L., Graff, C., Bird, T.D., Keene, C.D., Seeley, W.W., DeGrado, W.F., and Prusiner, S.B. (2019) Aβ and tau prion-like activities decline with longevity in the Alzheimer's disease human brain. Sci. Transl. Med. *11*, eaat8462, 1–13.

554) Krejciova, Z., Carlson, G.A., Giles, K., and Prusiner, S.B. (2019) Replication of multiple system atrophy prions in primary astrocyte cultures from transgenic mice expressing human α-synuclein. Acta Neuropathol. *137*, 437–454.

555) Woerman, A.L., Oehler, A., Kazmi, S.A., Lee, J., Halliday, G.M., Middleton, L.T., Gentleman, S.M., Mordes, D.A., Spina, S., Grinberg, L.T., Olson, S.H., and Prusiner, S.B. (2019) Multiple system atrophy prions retain strain specificity after serial propagation in two different Tg(*SNCA**A53T) mouse lines. Acta Neuropathol. *137*, 437–454.

556) Grandjean, J.-M.M., Jiu, A.Y., West, J.W., Aoyagi, A., Droege, D.G., Elepano, M., Hirasawa, M., Hirouchi, M., Murakami, R., Lee, J., Sasaki, K., Hirano, S., Ohyama, T., Tang, B.C., Vaz, R.J., Inoue, M., Olson, S.H., Prusiner, S.B., Conrad, J., Paras, N.A. (2020). Discovery of 4-piperazine isoquinoline derivatives as potent and brain-permeable tau prion inhibitors with CDK8 activity. ACS Med. Chem. Lett. *11*, 127–132.

557) Woerman, A.L., Patel, S., Kazmi, S.A., Oehler, A., Lee, J., Mordes, D.A., Olson, S.H., Prusiner, S.B. (2020). Kinetics of α-synuclein prions preceding neuropathological inclusions in multiple system atrophy. PLOS Pathog. *16*, e1008222, 1–18.

558) Lozano, C. Ramirez, C. Sin, N. Viart, H. M.-F. Prusiner, S. B. Paras, N. A. and Conrad, J. (2021) Silver benzoate facilitates the copper-catalyzed C–N coupling of iodoazoles with aromatic nitrogen heterocycles. ACS Omega *6*, 9804–9812.

559) Lester, E. Ooi, F. Bakkar, N. Ayers, J. Woerman, A. L. Wheeler, J. Bowser, R. Carlson, G. A. Prusiner, S. B. and Parker, R. (2021) Tau aggregates are RNA-protein assemblies that mislocalize multiple nuclear speckle components. Neuron *109*, 1675–1691.

560) Nguyen, T. H. O'Brien, C. J. Tran, M. L. N. Olson, S. H. Settineri, N. S. Prusiner, S. B. Paras, N. A. and Conrad, J. (2021) Water-Soluble Iridium Photoredox Catalyst for the Trifluoromethylation of Biomolecule Substrates in Phosphate Buffered Saline Solvent. Org Lett *23*, 3823-3827.

561) Ayers, J. I. Lee, J. Monteiro, O. Woerman, A. L. Lazar, A. A. Condello, C. Paras, N. A. and Prusiner, S. B. (In press) Different α-Synuclein Prion Strains Cause Dementia with Lewy Bodies and Multiple System Atrophy. Proc. Natl. Acad. Sci. U.S.A.

Single Author Books

1) Prusiner, S. B., *Madness and Memory: The Discovery of Prions—a New Biological Principle of Disease*, Yale University Press, New Haven, Conneticut, 344 pages, 2014. *Paperback edition 2015. Audiobook edition 2016.*

Edited Books

1) Prusiner, S., Stadtman, E. R. (eds.): *The Enzymes of Glutamine Metabolism*. Academic Press, New York, New York, 625 pages, 1973.

2) Prusiner, S. B., Hadlow, W. J. (eds.): *Slow Transmissible Diseases of the Nervous System, Vol. 1, Clinical, Epidemiological, Genetic, and Pathological Aspects of the Spongiform Encephalopathies*. Academic Press, New York, New York, 469 pages, 1979.

3) Prusiner, S. B., Hadlow, W. J. (eds.): *Slow Transmissible Diseases of the Nervous System, Vol. 2, Pathogenesis, Immunology, Virology, and Molecular Biology of the Spongiform Encephalopathies.* Academic Press, New York, New York, 524 pages, 1979.

4) Prusiner, S. B., McKinley, M. P. (eds.): *Prions – Novel Infectious Pathogens Causing Scrapie and Creutzfeldt-Jakob Disease.* Academic Press, Orlando, 534 pages, 1987.

5) Prusiner, S. B., Collinge, J., Powell, J., Anderton, B. (eds.): *Prion Diseases of Humans and Animals.* Ellis Horwood, London, England, 583 pages, 1992.

6) Rosenberg, R., Prusiner, S. B., DiMauro, S., Barchi, R., Kunkel, L. (eds.): *The Molecular and Genetic Basis of Neurological Disease.* Butterworth-Heinemann, Stoneham, Massachusetts, 1023 pages, 1993. Prusiner, S. B. (ed.): Prions Prions Prions. Current Topics in Microbiology and Immunology, Vol. 207, Springer-Verlag, Berlin-Heidelberg, 163 pages, 1996.

7) Rosenberg, R. N., Prusiner, S. B., DiMauro, S., Barchi, R. L. (eds.): *The Molecular and Genetic Basis of Neurological Disease*, Second Edition. Butterworth-Heinemann, Stoneham, Massachusetts, 1430 pages, 1997.

8) Rosenberg, R. N., Prusiner, S. B., DiMauro, S., Barchi, R. L. (eds.): *Clinical Companion to The Molecular and Genetic Basis of Neurological Disease*, Second Edition. Butterworth-Heinemann, Stoneham, Massachusetts, 318 pages, 1998.

9) Prusiner, S. B. (ed.): *Prion Biology and Diseases*. Cold Spring Harbor Laboratory Press, Cold Spring Harbor, New York, 794 pages, 1999.

10) Rosenberg, R. N., Prusiner, S. B., DiMauro, S., Barchi, R. L., Nestler, E. J. (eds.): *The Molecular and Genetic Basis of Neurologic and Psychiatric Disease*, Third Edition. Butterworth-Heinemann, Stoneham, Massachusetts, 844 pages, 2003.

11) Prusiner, S. B. (ed.): *Prion Biology and Diseases*, Second Edition. Cold Spring Harbor Laboratory Press, Cold Spring Harbor, New York, 1050 pages, 2004.

12) Prusiner, S. B. (ed.) Prion Biology. Cold Spring Harbor Laboratory Press, Cold Spring Harbor, NY, 456 pages, 2017.

13) Prusiner, S. B. (ed.): Prion Diseases. Cold Spring Harbor Laboratory Press, Cold Spring Harbor, New York, 677 pages, 2017.

Special Issues of Journals

1) Prusiner, S. B., DeArmond, S. J. (eds.): Prions. Brain Pathol. 5, 1995.

2) Prusiner, S. B. (ed.): Molecular Biology and Genetics of Prions. Semin. Virol. 7, 1996.

Journal Reviews (1980 – present): Total of 101

Book Chapters (1970 – present): Total of 225

Abstracts (1964 – present): Total of 729

Editorials, Letters and Other Publications (1977 – present): Total of 75

PATENTS

U.S. Patents Issued

Prusiner, S. B., Telling, G. C., Scott, M. R.: Method for Detecting Prions in a Sample and Transgenic Animal Used for Same. United States Patent Number 5,565,186. Issued October 15, 1996.

Prusiner, S. B., Kaneko, K., Cohen, F. E.: Formation and Use of Prion Protein (PrP) Complexes. United States Patent Number 5,750,361. Issued May 12, 1998.

Prusiner, S. B., Telling, G. C., Scott, M. R.: Method for Detecting Prions in a Sample and Transgenic Animal Used for Same. United States Patent Number 5,763,740. Issued June 9, 1998.

Prusiner, S. B., Telling, G. C., Scott, M. R.: Method for Detecting Prions in a Sample and Transgenic Animal Used for Same. United States Patent Number 5,789,655. Issued August 4, 1998.

Prusiner, S. B., Scott, M. R., Telling, G. C.: Detecting Prions in a Sample and Prion Preparation and Transgenic Animal Used for Same. United States Patent Number 5,792,901. Issued August 11, 1998.

Prusiner, S. B., Cohen, F. E., Muramoto, T.: A Soluble Form of PrP^{Sc} Which Is Insoluble in Native Form. United States Patent Number 5,834,593. Issued November 11, 1998.

Prusiner, S. B., Williamson, A., Burton, D. R.: Antibodies Specific for Native PrP^{Sc}. United States Patent Number 5,846,533. Issued December 8, 1998.

Prusiner, S. B., Safar, J.: Assay for a Disease Related Conformation of a Protein. United States Patent Number 5,891,641. Issued April 6, 1999.

Prusiner, S. B., Scott, M. R., Telling, G.: Method of Detecting Prions in a Sample and Transgenic Animal Used for Same. United States Patent Number 5,908,969. Issued June 1, 1999.

Prusiner, S. B., Cohen, F. E., James, T. L., Kaneko, K.: Nucleic Acid Encoding Prion Protein Variant. United States Patent Number 5,962,669. Issued October 5, 1999.

Prusiner, S. B. and Safar, J. G.: Process for Concentrating Protein with Disease-Related Conformation. United States Patent Number 5,977,324. Issued on November 2, 1999.

Prusiner, S. B., Scott, M. R., Telling, G.: Detecting Cow, Sheep and Human Prions in a Sample and Transgenic Mice Used for Same. United States Patent Number 6,008,435. Issued December 28, 1999.

Prusiner, S. B.: Prion Protein Standard and Method of Making Same. United States Patent Number 6,020,537. Issued February 1, 2000.

Prusiner, S. B., Telling, G. C., Cohen, F. E., Scott, M. R.: Transgenic Animals Expressing Artificial Epitope-tagged Proteins. United States Patent Number 6,150,583. Issued November 21, 2000.

Prusiner, S. B., Safar, J. G.: Method of Concentrating Prion Proteins in Blood Samples. United States Patent Number 6,166,187. Issued December 26, 2000.

Prusiner, S. B., Supattapone, S., Scott, M.: Clearance and Inhibition of Conformationally Altered Proteins. United States Patent Number 6,214,366 B1. Issued April 10, 2001.

Prusiner, S.B., Safar, J.: Assay for Disease Related Conformation of a Protein and Isolating Same. United States Patent Number 6,214,565 B1. Issued April 10, 2001.
Prusiner, S.B., Safar, J.: Removal of Prions from Blood, Plasma and Other Liquids. United States Patent Number 6,221,614 B1. Issued April 24, 2001.

Prusiner, S.B., Tremblay, P., Moore, R., Westaway, D., Hood, L.E., Lee, I.: PrP-Like Gene. United States Patent Number 6,277,970 B1. Issued August 21, 2001.

Prusiner, S.B., Williamson, A., Burton, D.R.: Antibodies Specific for Native PrP^{Sc}. United States Patent Number 6,290,954 B1. Issued September 18, 2001.

Prusiner, S.B., Supattapone, S., Scott, M.R.: Method of Sterilizing. United States Patent Number 6,322,802 B1. Issued November 27, 2001.

Prusiner, S.B., Supattapone, S., Scott, M.R.: Food Additives Which Affect Conformationally Altered Proteins. United States Patent Number 6,331,296 B1. Issued December 18, 2001.

Prusiner, S.B., Cohen, F.E., James, T.L., Kaneko, K.: Inhibitors of Prion Formation. United States Patent Number 6,365,359 B1. Issued April 2, 2002.

Prusiner, S.B., Williamson, R.A., Burton, D.R.: Antibodies Specific for Native PrP^{Sc}. United States Patent Number 6,372,214 B1. Issued April 16, 2002.

Prusiner, S.B., Safar, J.G.: Assay for Disease Related Conformation of a Protein and Isolating Same. United States Patent Number 6,406,864 B2. Issued June 18, 2002.

Prusiner, S.B., Supattapone, S., Scott, M.R.: Assay for Compounds Which Affect Conformationally Altered Proteins. United States Patent Number 6,419,916 B1. Issued July 16, 2002.

Prusiner, S.B., Supattapone, S., Scott, M.R.: Method of Sterilizing. United States Patent Number 6,517,855. Issued February 11, 2003.

Prusiner, S.B., Safar, J., Williamson, R. A., Burton, D.R.: Antibodies Specific for Ungulate PrP. United States Patent Number 6,537,548 B1. Issued March 25, 2003.

Prusiner, S.B., Williamson, A., Burton, D.R.: Antibodies Specific for Native PrP^{Sc}. United States Patent Number 6,562,341 B2. Issued May 13, 2003.

Prusiner, S.B., Telling, G.C., Cohen, F.C., Scott, M.R.: Recombinant Construct Encoding Epitope Tagged PrP Protein. United States Patent Number 6,602,672 B1. Issued August 5, 2003.

Prusiner, S.B., Safar, J., Cohen, F.C.: Assay for Specific Strains of Multiple Disease Related Conformations of a Protein. United States Patent Number 6,617, 119 B2. Issued September 9, 2003.

Prusiner, S.B., Safar, J.: Method for Detecting Prions. United States Patent Number 6,620,629 B1. Issued September 16, 2003.

Prusiner, S.B., Safar, J.G.: Assay for Disease Related Conformation of a Protein and Isolating Same. United States Patent Number 6,677,125. Issued January 13, 2004.

Prusiner, S. B., Supattapone, S.: Antiseptic Compositions for Inactivating Prions. United States Patent Number 6,719,988. Issued April 13, 2004.

Prusiner, S. B., Supattapone, S.: Sodium Dodecyl Sulfate Compositions for Inactivating Prions. United States Patent Number 6,720,355. Issued April 13, 2004.

Prusiner, S. B., Korth, C.: Models of Prion Disease. United States Patent Number 6,767,712. Issued July 27, 2004.

Prusiner, S. B.: Somatic Cells with Ablated PrP Gene and Methods of Use. United States Patent Number 6,797,495. Issued September 28, 2004.

Prusiner, S.B., Williamson, R.A., Burton, D.R.: Antibodies specific for native PrP^{Sc}. United States Patent Number 6,858,397. Issued February 22, 2005.

Prusiner, S.B., Safar, J.G.: Method of Preparing Cow Brain Homogenate. United States Patent Number 6,875,577. Issued April 5, 2005.

Prusiner, S.B., Safar, J.G.: Removal of Prions from Blood, Plasma, and Other Liquids. United States Patent Number 6,916,419. Issued July 12, 2005.

Prusiner, S.B.: Prion Protein Standard and Method of Making Same. United States Patent Number 6,962,975. Issued November 8, 2005.

Prusiner, S.B., Williamson, R. A., Burton, D. R.: Antibodies specific for native $PrP^{Sc.}$ United States Patent Number 7,052,675. Issued May 30, 2006.

Prusiner, S.B., Safar, J.G.: Method of preparing cow brain homogenate. United States Patent Number 7,087,213. Issued August 8, 2006.

Prusiner, S.B., Safar, J.G., Williamson, R. A., Burton, D. R.: Antibodies specific for ungulate PrP. United States Patent Number 7,094,553. Issued August 22, 2006.

Prusiner, S.B., Safar, J.G.: Method of concentrating proteins from serum. United States Patent Number 7,151,000. Issued December 19, 2006.

Prusiner, S.B., Safar, J.G.: Method of preparing cow brain homogenate. United States Patent Number 7,163,798. Issued January 16, 2007.

Prusiner, S.B.: Somatic cells with ablated PrP gene and methods of use. United States Patent Number 7,163,806. Issued January 16, 2007.

Prusiner, S.B., Supattapone, S.: Sodium dodecyl sulfate compositions for inactivating prions. United States Patent Number 7,226,609. Issued June 5, 2007.

Prusiner, S.B., Supattapone, S.: Sodium dodecyl sulfate compositions for inactivating prions. United States Patent Number 7,307,103. Issued December 11, 2007.

Prusiner, S.B., Supattapone, S.: Sodium dodecyl sulfate compositions for inactivating prions. United States Patent Number 7,307,103. Issued December 11, 2007.

Prusiner, S.B., Price, J.C., Ghaemmaghami, S., Burlingame, A.L., Guan, S.: Isotopic labeling for the measurement of global protein levels and turnover in vivo, United States Patent Application Number 13/704,498. – Issued September 24, 2014.

U.S. Patents Pending

Prusiner, S.B., Serban, A., Ana, V., Safar, J., Stanker, L.: Antibodies specific for human and bovine PrP, United States Patent Application Number 11/817,488.

Prusiner, S.B., Korth, C., May, B.: Optically active compounds clearing malformed proteins, United States Patent Number 10/478,560.

Prusiner, S.B., Price, J.C., Ghaemmaghami, S., Burlingame, A.L., Guan, S.: Isotopic labeling for the measurement of global protein levels and turnover in vivo, United States Patent Application Number 13/704,498.

International Patents Issued

Prusiner, S.B., Williamson, R.A., Burton, D.R.: Antibodies Specific for Native PrP^{Sc}. Mexican Patent Number 204412. Issued September 27, 2001.

Prusiner, S. B., Scott, M. R., Telling G. C.: Detecting Cow, Sheep and Pig Prions in a Sample and Transgenic Animal Used for Same. Australian Patent Number 752933. Issued January 16, 2003.

Prusiner, S.B., Safar, J.G., Cohen, F. E.: Assay for Specific Strains of Multiple Disease Related Conformations of a Protein. Australian Patent Number 26602/99. Issued February 6, 2003.

Prusiner, S.B., Williamson, R.A., Burton, D.R.: Antibodies Specific for Native PrP^{Sc}. Canadian Patent Number 2,231,409. Issued February 11, 2003.

Prusiner, S.B., Safar, J.G.: Process for Concentrating Protein with Disease-related Conformation. Australian Patent Number 761710. Issued September 18, 2003.

Prusiner, S.B., Scott, M.R., Supattapone, S.: Method of Sterilizing. New Zealand Patent Number 515607. Issued November 3, 2003.

Prusiner, S.B.: Prion Protein Standard and Method of Making Same. New Zealand Patent Number 511482. Issued December 8, 2003.

Prusiner, S.B., Safar, J.G.: Assay for Disease Related Conformation of a Protein. Australian Patent Number 764888. Issued January 8, 2004.

Prusiner, S.B., Safar, J.G.: Assay for Disease Related Conformation of a Protein. Australian Patent Number 765939. Issued January 15, 2004.

Prusiner, S.B., Safar, J.G.: Removal of Prions from Blood, Plasma and Other Liquids. Australian Patent Number 768032. Issued March 11, 2004.

Prusiner, S.B.: Somatic Cells with Ablated PrP Gene and Methods of Use. Australian Patent Number 767893. Issued March 18, 2004.

Prusiner, S.B., Safar, J.G.: Blood Serum Sample Prepared for Isolation of Prions and PrP^{Sc}. Australian Patent Number 768654. Issued April 1, 2004.

Prusiner, S.B.: Prion Protein Standard and Method of Making Same. Australian Patent Number 768840. Issued April 22, 2004.

Prusiner, S.B., Supattapone, S., Scott, M.R.: Method of Sterilizing. Australian Patent Number 771547. Issued July 8, 2004.

Prusiner, S.B., Safar, J.G.: Removal of Prions from Blood, Plasma and Other Liquids. Chinese Patent Number ZL 99815767.8. Issued December 29, 2004.

Prusiner, S.B., Safar, J.G.: Assay for a Prion Protein in its Disease Related Conformation. European Patent Number 1119773 (German Patent Number 69926248.8–08). Issued July 20, 2005.

Prusiner, S.B., Safar, J.: Assay for disease related conformation of a protein. Japanese Patent Number 3774236. Issued February 24, 2006.

Prusiner, S.B., Burton, D.R., Safar, J., Williamson, A.: Antibodies specific for ungulate PrP. Australia Patent Number 2001275977. Issued February 1, 2007.

International Patents Pending

DeArmond, S., Prusiner, S.: Combined gamma-secretase inhibitor and phenothiazine derivative treatment of neurodegenerative diseases, Serial Number US2007/018857, Filed: 08/27/2007, Publication Number WO2008/027345, Published 03/06/2008.

Woerman, A. L., Olson, S. H., Aoyagi, A., Prusiner, S. B., Patel, S., and Kazmi, S.: Modified cell line and method of determining tauopathies, Filed March 28, 2017; International Patent Number, WO2017/172764A1, Published October 5, 2017.