

JEFFERY W. KELLY

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Birthday: 8/23/60, born in Medina, N.Y. **Office:** 858-784-9880

Administrative Laboratory Manager: Colleen Fearn, Ph.D; **Fax:** 858-784-9610

Education:

B.S. Chemistry, (1982) State University of New York College at Fredonia (Tom Harris)
Ph.D. Organic Chemistry, (1986) University of North Carolina (Slayton A. Evans, Jr.)
NIH Postdoctoral Training, (1986-1989) The Rockefeller University (E. Thomas Kaiser)

Academic Experience:

Chairman, Board of Trustees, The Skaggs Institute for Research, 2008-2010
Chairman, Molecular and Experimental Medicine, Scripps, 2008-
Dean of Graduate Studies, The Scripps Research Institute 2000-2008
Vice President, Academic Affairs, The Scripps Research Institute 2000-2006
Lita Annenberg Hazen Professor of Chemistry, Department of Chemistry and the
Skaggs Institute for Chemical Biology, The Scripps Research Institute, 1997-
President, Protein Society 2005-2007 (elected position)
Professor of Chemistry, Texas A&M University, 1997
Visiting Investigator, Memorial Sloan-Kettering Institute, Summer 1996
Associate Professor of Chemistry, Texas A&M University, 1995-1997
Assistant Professor of Chemistry, Texas A&M University, 1989-1995

Translational Medicine Experience:

Co-founder of FoldR_x Pharmaceuticals, Boston, MA 2003-Enabled Vyndaqel (Tafamidis)
European Drug Agency Approval in 2011 and Japanese approval (2013). This
compound was discovered by the Kelly Laboratory at Scripps to treat Familial
Amyloid Polyneuropathy / Cardiomyopathy, FDA approval expected in 2018. First
drug to treat the underlying etiology of any human amyloid disease and the first
pharmacological evidence supporting the amyloid hypothesis—the notion that the
process of amyloidogenesis, but not amyloid fibrils themselves, causes post-mitotic
tissue degeneration. FoldR_x was acquired by Pfizer in October, 2010.
Co-founder Proteostasis Therapeutics, Boston 2007- (with Morimoto, Dillin, Finley, King)
Developing Proteostasis Regulators, compounds that restore or enhance cellular
protein homeostasis to ameliorate folding / aggregation diseases.
Repurposing: enabled positive diflunisal clinical trial for familial amyloid polyneuropathy,
based on the Kelly Lab discovery that diflunisal kinetically stabilizes transthyretin.

Board Service:

Board of Directors and SAB, Yumanity Therapeutics, Neurodegenerative Diseases, 2015-
Board of Directors and SAB, Proteostasis Therapeutics, Cystic Fibrosis, etc. 2008-
Chair, Therapeutic Advisory Panel, Pfizer, Orphan and Rare Diseases, 2012-2015
Pfizer Therapeutic Advisory Panel, Orphan and Rare Diseases, 2015-
Chair, Scientific Advisory Board, Viewpoint Therapeutics, Cataract Treatment, 2015-
Scientific Advisory Board, Neuropore Therapies, Alzheimer's disease 2014-
Scientific Advisory Board, Lysosomal Therapeutics, Parkinson's disease 2014-
Board of Directors, Amyloidosis Research Consortium 2015-

Awards:

Jacob Heskel Gabbay Award in Biotechnology and Medicine, 2016
American Academy of Arts and Sciences, Elected member, 2016
Royal Society of Chemistry Jeremy Knowles Award, 2016
Biopolymers Murray Goodman Memorial Prize, 2012
American Chemical Society Ralph F. Hirschmann Award in Peptide Chemistry, 2012
The Protein Society Emil Thomas Kaiser Award, 2011
The American Peptide Society Rao Makineni Lectureship (Award), 2011
The American Peptide Society Vincent du Vigneaud Award, 2008
National Institutes of Health Merit Award, 2006
American Chemical Society Arthur C. Cope Scholar Award, 2001
State University of New York at Fredonia Alumni Distinguished Achievement Award, 2000
State University of New York at Fredonia Chemistry Department Alumni Award, 2000
The Protein Society–Dupont Young Investigator Award, 1999
The Biophysical Society National Lecturer (Award), 1999
Texas A&M University Honors Program Teacher / Scholar Award, 1994-1995
Camille Dreyfus Teacher-Scholar Award, 1994
Searle Scholar Award, 1991-1994
NIH Postdoctoral Fellowship, The Rockefeller University, 1987-1989
American Chemical Society Rohm and Haas Organic Division Fellowship, 1985-1986
American Chemical Society North Carolina Centennial Scholarship, 1986
American Institutes of Chemists Outstanding Senior in Chemistry, 1982

Partial List of Academic Service:

SUNY Fredonia College Foundation Board of Directors, 2010-
Scientific Advisory Board, Gladstone's Center for Alzheimer's Research, UCSF, 2012-
Scientific Advisory Board, Hope Center for Neurological Disorders, Wash. Univ. 2012-
Scientific Advisory Board, Proteostasis Therapeutics, 2005-
NIH National Eye Institute, Nanomedicine Medical Advisory Board, 2007-
Elan Pharmaceuticals Neurodegenerative Disease Advisory Board 2010-
Editorial Advisory Board, *Biochemistry*, 2010-2012.
Genomics Institute of the Novartis Foundation Outside Review Team, 2009
Harvard Medical School, Therapeutics Advisory Group 2008
Chair, UCSF Chemical Biology Graduate Program Review 2008
Board of Directors, Grossman Center for Memory Research and Care, U. Minn. 2007-
NIH Special emphasis panel in cell biology 2006
Board Member, "Prion" Journal 2006-
Scientific Advisor, Amyloidosis Research Foundation 2006-
Co-organizer, Banbury Conference on "Functional & Pathologic Amyloids" Spring 2007
Facilitated FDA and NIH funded Diflunisal clinical trial on Amyloid Polyneuropathy
Board Member, International Journal of Peptide Research and Therapeutics, 2004-
Co-organizer, Joint Keystone Symposia on Alzheimer's Disease: Genes, Cellular Pathways and Therapies and Protein Misfolding with Lindquist & Tanzi and Lee 2006.
Chairman, NIH Roadmap Initiative Screening Assay Dev. Study Section 2004
American Chemical Society Committee on Professional Training 2003-4
Meeting Co-chair-American Peptide Symposium, 2005 (with Tom Muir)
The Celiac Sprue Research Foundation-Scientific Advisory Board Member ,2002-

Partial List of Academic Service:

Vanderbilt Institute of Chemical Biology- External Advisory Committee, 2002-2006
 Scientific Advisory Board, Provid Pharmaceuticals Inc., 2002-
 Rockefeller University Graduate Program Review 2001
 Hereditary Disease Foundation Scientific Advisory Board 2001-2006
 Chairman, NIH Bioorganic and Natural Products Study Section 1998-2000
 Chairman, NIH Special Study Section on Protein Structure, Function and Aging, 1999
 Protein Science, Editorial Advisory Board 1999-
 Bioorganic and Medicinal Chemistry-Board of Consulting Editors 2004-2009
 Biopolymers, Editorial Board and Advisory Board 2001-, 2002-
 NIH-NINDS Planning Committee on Neurodegenerative Diseases 1998
 Co-Chair, Chemistry and Biology of Peptides Gordon Conference 2002
 Co-Chair, "Peptide Chemistry as a Life Science Symposium" at PacificChem 2000 Mtg.
 Advisory Board-International Symposium on Human Amyloid Diseases-1999
 Co-Organizer, Protein Society Meeting, Boston, 1999
 NIH Bioorganic and Natural Products Study Section Member-October 1996-1998
 Volume Editor-Comprehensive Natural Products Chemistry, Vol 4, Amino Acids, Peptides
 Organizing Committee-Self Assembling Peptides in Biology, Medicine Crete, Greece 1999
 Program Committee "16th American Peptide Symposium, Minneapolis 1999
 Scientific Advisory Board VIII International Symposium on Amyloidosis, 1998
 NIH Aging Study Section 1997, 1998, 1999-Ad Hoc
 Co-organizer NIH-National Institute on Aging Aging and Disease Workshop
 External Reviewer, Veterans Association Intramural Program
 Alzheimer's Association Study Section, 1996, 1997
 NIH Bioorganic Chemistry and Related Sciences Special Study Section, 1996
 NIH Medical Biochemistry Study Section Ad Hoc Reviewer, September 1996
 Co-Founder FASEB Summer Research Conference On "Amyloid and Other Abnormal
 Protein Assembly Processes"-1995 (with Ron Wetzel)
 National Institutes of Health Bioorganic and Natural Products Study Section, Ad Hoc 1995
 Editorial Board-*Journal of Amyloidosis*, 1994-
 Scientific Advisory Board- *International Symposium on FAP and Related Diseases*
 Advisory Board-Peptide Synthesis Research Committee of the Association of
 Consultant-Parke Davis 1996-2001
 Consultant-Praecis Pharmaceuticals 1996-2000
 Consultant-Hoffmann-LaRoche Allergy and Inflammation-1996-1997

Lecture Courses Taught:

Texas A&M University

CH 481	Senior Seminar, 1996
CH 646	Mechanistic Organic Chemistry, 1989, 1990, 1991
CH 681	Organic Seminar 1989, 1990, 1991
CH 653	The Chemistry of Peptides and Proteins, 1990
CH 446	Advanced Organic Chemistry (Undergraduate), 1991
CH 672	Bioorganic (Enzyme) Reaction Mechanisms, 1992
CH 647 / 465	Spectroscopic Identification of Organic Compounds, 1993-96
CH 689	Special Topics in Medicinal Chemistry Spring, 1997
CH 234	Majors Organic Laboratory Course With Lecture (3h), 90,92,94-95

Lecture Courses Taught-continued:

Scripps Research Institute Physical Organic Chemistry (1998-2000 with Professor Rebek)
 Structure and Chemistry (2 -3 lectures every year)
 Chemical Biology 1 (responsible for entire course every third year)
 Organic Spectroscopy (responsible for one lecture of course)
 Bioorganic Chemistry II (responsible for entire course **2003**)
 Chemical Biology (taught every other year)

Partial list of Scripps Service:

Raised \$ 110,000 as Chairman in year 2 to hire an Asst. Prof. at Scripps
 Raised \$ 400,000 as Chairman in year 2 to hire Sandra Encalada at Scripps
 Raised \$ 1.5 mm as Chairman in year 1 to hire Luke Wiseman at Scripps
 Raised \$ 1.0 mm as Vice President in 2005 to use at TSRI as needed
 Responsible for Reaccreditation of TSRI as an academic institution 2005-2008
 Dean of Graduate Studies, 2000-2008, responsible integrating chemistry and biology
 programs into a single program and improving the top 10 ranking of this program
 Chair-Graduate Recruiting-Chemistry Program 1998-2000
 Scripps-Novartis Joint Scientific Council 1997-2006
 Chair-Reaccreditation Curriculum Subcommittee, both Graduate Programs at Scripps 1997
 Advisory Board-Harold L. Dorris Neurological Research Center
 PI of shared instrumentation proposal that successfully equipped walk-up MS facility

39 of 86 Individuals Educated in the Kelly Laboratory who are now in tenured or tenure-track professorial positions:

Wilfredo Colon Graduate Student	Ph.D.	Associate Professor Department of Chemistry Rensselaer Polytechnic Institute, NY, USA
Joel Schneider Graduate Student	Ph.D.	Professor-Chemical Biology, Deputy Director National Institutes of Health, NCI. Formerly, Prof. Univ. Delaware
Hilal Lashuel Graduate Student	Ph.D.	Professor and Director Ecole Polytechnique Federal (ETH) Lausanne, School of Life Sciences, Lausanne, Switzerland
Jose M. Rivera Post-doctoral Fellow	Ph.D.	Associate Professor Department of Chemistry University of Puerto Rico, Rio Piedras
Evan T. Powers Post-doctoral Fellow	Ph.D.	Associate Professor Department of Chemistry The Scripps Research Institute, CA, USA
Yoshiki Sekijima Post-doctoral Fellow	M.D., Ph.D.	Professor Department of Neurology & Rheumatology Shinshu University School of Medicine, Japan
Per Hammarstrom Post-doctoral Fellow	Ph.D.	Professor Department of Biochemistry Linkoping University, Sweden
Shu-Li You Post-doctoral Fellow	Ph.D.	Professor State Key Laboratory of Organometallic Chem. Shanghai Institute of Organic Chemistry, China

Selected Individuals Educated in the Kelly Laboratory of > 90 trainees-cont-

Jianmin Gao Post-doctoral Fellow	Ph.D.	Associate Professor Department of Chemistry Boston College, MA, USA
Laura Segatori Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemical Engineering Rice University, TX, USA
Qinghai Zhang Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Molecular Biology The Scripps Research Institute, CA, USA
Tingwei Mu Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Physiology and Biophysics Case Western Reserve Univ., School of Medicine
Deguo Du Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry Florida Atlantic University
Nora Green Post-doctoral Fellow	Ph.D.	Associate Professor Department of Chemistry Randolf-Macon College, USA
Lyudmila Bazhenova Post-doctoral Fellow	M.D.	Assistant Professor Department of Oncology University of California at San Diego, USA
Daryl Bosco Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Neurology Univ. of Massachusetts Medical School, USA
R. Luke Wiseman Graduate Student	Ph.D.	Assistant Professor Dept. of Molecular and Experimental Medicine The Scripps Research Institute
Jan Bieschke Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Biomedical Engineering Washington Univ. St. Louis
Songpon Deechongkit Graduate Student	Ph.D.	Professor Chulabhorn Research Institute Thailand
Kenji Usui Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Nanotechnology Konan University, Japan
Praveen Nekkar Post-doctoral Fellow	Ph.D.	Assistant Professor Pharmacy School University of Waterloo, Canada
Amelia Fuller Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry Santa Clara College, CA, USA
Sarah Siegel Graduate Student	Ph.D.	Assistant Professor Department of Chemistry Gonzaga University, Spokane, WA, USA
Paul Baures Post-doctoral Fellow	Ph.D.	Associate Professor Department of Chemistry University of Tulsa, USA

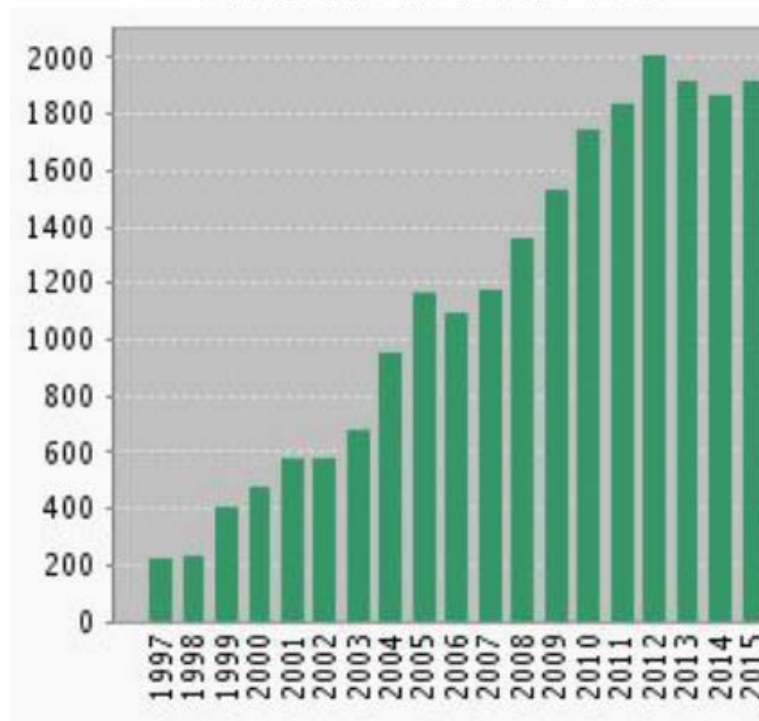
Selected Individuals Educated in the Kelly Laboratory of > 80 trainees-cont-

Sandra McCutchen Graduate Student	Ph.D.	Professor of Biosciences Lawrence Livermore National Laboratory
Sungwook Choi Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry Chungnam National University, South Korea
Joshua L. Price Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry Brigham Young University
Derrick Suk Tong Ong Graduate Student	Ph.D.	Assistant Professor Faculty of Medicine National University of Singapore
Steven Bourgault Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry University of Quebec at Montreal
Fernando L. Palhano Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Biochemistry Universidade Federal do Rio de Janeiro, Brazil
Matthew Shoulders Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry Massachusetts Institute of Technology
Jiyong Lee Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry University of Texas at Dallas
Doug Fowler Graduate Student	Ph.D.	Assistant Professor Department of Genome Sciences and Biochemistry University of Washington Seattle
Steven M. Johnson Graduate Student	Ph.D.	Assistant Professor Department of Biochemistry and Molecular Biology Indiana University School of Medicine
John D. Hulleman Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Ophthalmology UT Southwestern School of Medicine
Lisa D. Ryno Graduate Student	Ph.D.	Assistant Professor Department of Chemistry Oberlin College
Christina B. Cooley Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry Trinity University
Joseph C. Genereux Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry University of California, Riverside
Xin Zhang Post-doctoral Fellow	Ph.D.	Assistant Professor Department of Chemistry Pennsylvania State University

Publication List–Jeffery W. Kelly:

ISI ***h-index*** = **83**, 83 publications with 83 or more citations
(> 67 papers with ≥ 100 citations, 23,000+ total citations)
Google Scholar ***h-index*** = **98**

Citations in Each Year



The latest 20 years are displayed.

Data from ISI database as of April 5, 2016

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- 320 Plate, L.; Paxman, R.J.; Wiseman, R.L.; Kelly, J.W. “Unfolded protein response: Modulating protein quality control” *eLife* **2016**;5:e18431.
- 319 Kurian, S.M.; Novias, M.; Whisenant, T.; Gelbart, T.; Buxbaum, J.N.; Kelly, J.W.; Coelho, T.; Salomon, D.R. “Peripheral Blood Cell Gene Expression Diagnostic for Identifying Symptomatic Transthyretin Amyloidosis Patients: Male and Female Specific Signatures” *Theranostics* **2016** 6, 1792-1809.
- 318 Hsu, C-H.; Park, S.; Mortenson, D.E.; Foley, B.L.; Wang, X.; Woods, R.J.; Case, D.A.; Powers, E.T.; Wong, C-H.; Dyson, H.J.; Kelly J.W. “The Dependence of

Carbohydrate–Aromatic Interaction Strengths on the Structure of the Carbohydrate” *J. Am. Chem. Soc.* **2016** 138, 7636-7648.

- 317 Plate, L.; Cooley, C.B.; Chen, J.J.; Paxman, R.J.; Gallagher, C.M.; Madoux, F.; Genereux, J.C.; Dobbs, W.; Garza, D.; Spicer, T.P.; Scampavia, L.; Brown, S.J.; Rosen, H.; Powers, E.T.; Walter, P.; Hodder, P.; Wiseman, R.L.; Kelly, J.W. “Small Molecule Proteostasis Regulators that Reprogram the ER to Reduce Extracellular Protein Aggregation” *eLife* **2016**;5;15550.
- 316 Chen, W.; Dong, J.; Plate, L.; Mortenson, D.E.; Brighty, G.J.; Li, S.; Liu, Y.; Galmozzi, A.; Lee, P.S.; Hulce, J.J.; Cravatt, B.F.; Saez, E.; Powers, E.T.; Wilson, I.A.; Sharpless, K.B.; Kelly, J.W. “Arylfluorosulfates Inactivate Intracellular Lipid Binding Protein(s) through Chemoselective SuFEx Reaction with a Binding-site Tyr Residue” *J. Am. Chem. Soc.* **2016** 138, 7353-7364.
- 315 Chen, W.; Kong, L.; Connelly, S.; Dendle, J.M.; Liu, Y.; Wilson, I.A.; Powers, E.T.; Kelly, J.W. “Stabilizing the C_{H2} domain of an Antibody by Engineering in an Enhanced Aromatic Sequon” *ACS Chem. Biol.* **2016** 11, 1852-1861.
- 314 Lim, K.H.; Dasari, A.K.R.; Hung, I. Gan, Z.; Kelly J.W.; Wemmer, D.E. “Structural Changes Associated with Transthyretin Misfolding and Amyloid Formation Revealed by Solution and Solid-State NMR” *Biochemistry* **2016** 55, 1941-1944.
- 313 Kapil, D.; Jager, M.; Nguyen, H.; Kelly, J.W.; Gruebele, M. “High Resolution mapping of the folding transition state of a WW domain” *J. Mol. Biol.* **2016** 428, 1617-1636.
- 312 Coelho, T.; Merlini, G.; Bulawa, C.E.; Fleming, J.A.; Judge, D.P.; Kelly, J.W.; Maurer, M.S.; Plante-Bordeneuve, V.; Labaudiniere, R.; Mundayat, R.; Riley, S.; Lombardo, I.; Huertas, P.; “Mechanism of Action and Clinical Application of Tafamidis in Hereditary Transthyretin Amyloidosis” *Neurology and Therapy* **2016** 5: 1. doi:10.1007/s40120-016-0040-x.
- 311 Baranczak, A.; Kelly, J.W. “A Current Pharmacologic Agent versus The Promise of Next Generation Therapeutics to Ameliorate Protein Misfolding and/or Aggregation Diseases” *Curr. Op. Chem. Biol.* **2016** 32 10-21.
- 310 Ankarcrona, M.; Winblad, B.; Monteiro, C.; Fearn, C.; Powers, E.T.; Johansson, J.; Westermark, G.T.; Presto, J.; Ericzon, B-G.; Kelly, J.W. “Current and Future Treatment of Amyloid Diseases” *J. Internal Medicine* **2016** 280 177-202.
- 309 Chen, W.; Dong, J.; Li, S.; Liu, Y.; Wang, Y.; Yoon, L.; Wu, P.; Sharpless, K.B.; Kelly, J.W. “Synthesis of Sulfotyrosine-Containing Peptides by Incorporating Fluorosulfated Tyrosine Using an Fmoc Solid-phase Strategy” *Angew. Chem. Int. Ed.* **2016** 55, 1835-1838.

- 308 Liu, Y.; Zhang, X.; Chen, W.; Tan, Y.; Kelly, J.W. "Fluorescence Turn-on Folding Sensor to Monitor Proteome Stress in Live Cells" *J. Am. Chem. Soc.* **2015** 137, 11303-11311.
- 307 Cho, Y.H.; Baranczak, A.; Helmke, S.; Teruya, S.; Horn, E.M.; Maurer, M.S.; Kelly, J.W. "Personalized Medicine Approach for Optimizing the Dose of Tafamidis to Potentially Ameliorate Wild-type Transthyretin Amyloidosis (Cardiomyopathy)" *Amyloid* **2015** 22, 175-180.
- 306 Baranczak, A.; Liu, Y.; Connelly, S.; Dan Du, W-G.; Greiner, E.R.; Genereux, J.C.; Wiseman, R.L.; Eisele, Y.S.; Bradbury, N.C.; Dong, J.; Noodleman, L.; Sharpless, K.B.; Wilson, I.A.; Encalada, S.E.; Kelly, J.W. "A Fluorogenic Aryl Fluorosulfate for Intraorganellar Transthyretin Imaging in Living Cells and in *Caenorhabditis elegans*" *J. Am. Chem. Soc.* **2015** 137, 7404-7414.
- 305 Murray, A.N.; Chen, W.; Antonopoulos, A.; Hanson, S.R.; Wiseman, R.L.; Dell, A.; Haslam, S.M.; Powers, D.L.; Powers, E.T.; Kelly, J.W. "Enhanced aromatic sequons increase oligosaccharyltransferase glycosylation efficiency and glycan homogeneity" *Chem. and Biol.* **2015** 22, 1052-1062.
- 304 Eisele, Y.S.; Monteiro, C.; Fearn, C.; Encalada, S.E.; Wiseman, R.L.; Powers, E.T.; Kelly, J.W. "Targeting Protein Aggregation to Ameliorate Degenerative Diseases" *Nature Rev. Drug Discovery* **2015** 14, 759-780.
- 303 Cho, Y.H.; Zhang, X.; Pobre, K.F.R.; Liu, Y.; Powers, D.L.; Kelly, J.W.; Gierasch, L.M. Powers, E.T. "Individual and Collective Contributions of Chaperoning and Degradation to Protein Homeostasis in *E. coli*" *Cell Reports* **2015** 11, 321-333.
- 302 Genereux, J.C.; Qu, S.; Zhou, M.; Cooley, C.B.; Ryno, L.M.; Madrazo, N.; Rainbolt, T.K.; Wang, S.; Shoulders, M.D.; Kaufman, R.J. Lasmezas, C.I.; Kelly, J.W.; Wiseman, R.L. "Unfolded Protein Response-induced ERdj3 Secretion Links ER Stress to Extracellular Proteostasis" *EMBO J.* **2015** 34, 4-19.
- 301 Hulleman, J.D.; Kelly, J.W. "Genetic Ablation of N-linked Glycosylation Reveals Two Key Folding Pathways for R345W Fibulin-3, a Protein Associated with Retinal Degeneration" *FASEB J.* **2015** 29, 565-575.
- 300 Liu, Y.; Zhang, X.; Tan, Y-L; Bhabha, G.; Ekiert, D.C.; Kipnis, Y.; Bjelic, S. Baker, D.; Kelly, J.W. "De novo Designed Enzymes as Small Molecule-regulated Fluorescence Imaging Tags and Fluorescent Reporters" *J. Am. Chem. Soc.* **2014** 136, 13102-13105.
- 299 Zhang, X.; Liu, Y.; Genereux, J.C.; Nolan, C.; Singh, M.; Kelly, J.W. "Heat Shock Response Transcriptional Program Enables High Yield and High-Quality Recombinant Protein Production in *Escherichia coli*" *ACS Chem. Biol.* **2014** 9, 1945-1949.

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- 297 Cooley, C.B.; Ryno, L.M.; Plate, L.; Morgan, G.J.; Hulleman, J.D.; Kelly, J.W.; Wiseman, L.R. "Unfolded Protein Response Activation Reduces Secretion and Extracellular Aggregation of Amyloidogenic Light Chain" *Proc. Natl. Acad. Sci. USA* **2014** 111, 13046-13051.
- 296 Hebert, D.N.; Lamriben, L.; Powers, E.T.; Kelly, J.W. "The Intrinsic and Extrinsic Effects of N-linked Glycans on Glycoproteostasis" *Nature Chem. Biol.* **2014** 10, 902-910.
- 295 Zhang, X.; Kelly, J.W. "Chaperonins Resculpt Folding Free Energy Landscapes to Avoid Kinetic Traps and Accelerate Protein Folding" *J. Mol. Biol.* **2014** 426, 2736-2738.
- 294 Tan, Y.L.; Genereux, J.C.; Pankow, S.; Aerts, J.M.F.G.; Yates, J.R. III; Kelly, J.W. "ERdj3 is an Endoplasmic Reticulum Degradation Factor for Mutant Glucocerebrosidase Variants Linked to Gaucher's Disease" *Chem. and Biol.* **2014** 21, 967-976.
- 293 Rappley, I.; Monteiro, C.; Novias, M.; Baranczak, A.; Solis, G.; Wiseman, R.L.; Helmke, S.; Maurer, M.S.; Coelho, T.; Powers, E.T.; Kelly, J.W. "Quantification of Transthyretin Kinetic Stability in Human Plasma Using Subunit Exchange" *Biochemistry* **2014** 53, 1993-2006.
- 292 Liu, Y.; Tan, Y.L.; Zhang, X. Bhabha, G.; Ekiert, D.C. Genereux, J.C.; Cho, Y.H.; Kipnis, Y.; Bjelic, S.; Baker, D.; Kelly, J.W. "Small Molecule Probes to Quantify the Functional Fraction of a Specific Protein in a Cell with Minimal Folding Equilibrium Shifts" *Proc. Natl. Acad. Sci USA*. **2014** 111, 4449-4454.
- 291 Berk, J.L.; Suhr, O.; Obici, L.; Sekijima, Y.; Zeldenrust, S.R.; Yamashita, T.; Hennehan, M.A.; Gorevic, P.D.; Litchy, W.J.; Wiseman, J.F.; Nordh, E.; Corato, M.; Lozza, A.; Cortese, A.; Robinson-Papp, J.; Colton, T.; Rybin, D.V.; Bisbee, A.B.; Ando, Y.; Ikeda, S.; Seldin, D.C.; Merlini, G.; Skinner, M.; Kelly, J.W.; Dyck, P.J. "Repurposing Diflunisal for Familial Amyloid Polyneuropathy—a Randomized Clinical Trial" *J. Am. Med. Assoc.* **2013** 310, 2658-2667.
- 290 Nunes, R.J.; de Oliveira, P.; Lages, A.; Becker, J.D.; Marcelino, P.; Barroso, E.; Kelly, J.W.; Quintas, A.; Constantino Rosa Santos, S. "Transthyretin Proteins Regulate Angiogenesis by Conferring Different Molecular Identities to Endothelial Cells" *J. Biol. Chem.* **2013** 288, 31752-31760.
- 289 Li, X.; Zhang, X.; Ladiwala, A.R.; Du, D.; Yadav, J.; Tessier, P.; Wright, P.; Kelly, J.W.; Buxbaum, J.N. "Mechanisms of transthyretin inhibition of Aβ aggregation in vitro" *J. Neuroscience* **2013** 33, 19423-19433

- 288 Suh, E.H.; Liu, Y.; Connelly, S.; Genereux, J.C.; Wilson, I.A.; Kelly, J.W. "Stilbene Vinyl Sulfonamides as Fluorogenic Sensors of and Traceless Covalent Kinetic Stabilizers of Transthyretin that Prevent Amyloidogenesis" *J. Am. Chem. Soc.* **2013** 135, 17869-17880.
- 287 Baranczak, A.; Connelly, S.P.; Liu, Y.; Choi, S.; Grimster, N.P.; Powers, E.T.; Wilson, I.A.; Kelly, J.W. "Fluorogenic Small Molecules Requiring Reaction with a Specific Protein to Create a Fluorescent Conjugate for Biological Imaging—What We Know and What We Need to Learn" *Biopolymers* **2013** 101, 485-495.
- 286 Coelho, T.; Maia, L.F.; Martins da Silva, A.; Cruz, M.W.; Planté-Bordeneuve, V.; Suhr, O.B.; Conceição, I.; Schmidt, H. H. J.; Trigo, P. Kelly, J.W.; Labaudiniere, R.; Chan, J., Packman, J.; Grogan, D.R. "Long-term Effects of Tafamidis for the Treatment of Transthyretin Familial Amyloid Polyneuropathy" *J. Neurology* **2013** 260, 2802-2814.
- 285 Myung, N.; Connelly, S.; Kim, B.; Park, S.J.; Wilson, I.A.; Kelly, J.W.; Choi, S. "Bifunctional Coumarin Derivatives That Inhibit Transthyretin Amyloidogenesis and Serve as Fluorescent Transthyretin Folding Sensors" *Chem Comm.* **2013** 49, 9188-9190.
- 284 Bannykh, S.I.; Balch, W.E.; Kelly, J.W.; Page, L.J.; Shelton, G.D. "Formation of Gelsolin Amyloid Fibrils in the Rough Endoplasmic reticulum of Skeletal Muscle in the Gelsolin Mouse Model of Inclusion Body Myositis: Comparative Analysis to Human Sporadic Inclusion Body Myositis" *Ultrastructural Pathology* **2013** 37, 304-311.
- 283 Chen, W; Enck, S.; Price, J.L.; Powers, D.L.; Powers, E.T.; Wong, C-H.; Dyson, H.J.; Kelly, J.W. "The Structural and Energetic Basis of Protein-Carbohydrate Interactions" *J. Am. Chem. Soc.* **2013** 135, 9877-9884.
- 282 Ryno, L.M.; Wiseman, R.L.; Kelly, J.W.; "Targeting Unfolded Protein Response Signaling Pathways to Ameliorate Protein Misfolding Diseases" *Curr. Op. Chem. Biol.* **2013** 17, 346-352.
- 281 Penchala, S.C.; Connelly, S.; Wang, Y. Zhao, L.; Baranczak, A.; Rappley, I.; Liedtke, M.; Park, M.S.; Powers, E.T.; Reixach, N.; Chan, W.K.; Wilson, I.A.; Kelly, J.W.; Graef, I.A.; Alhamadsheh, M.M. "AG10 Inhibits Amyloidogenesis and Cellular Toxicity of the Familial Amyloid Cardiomyopathy Associated V122I Transthyretin" *Proc. Natl. Acad. Sci USA.* **2013** 110, 9992-9997.
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Patents

- 1 Transthyretin Antibodies: amyloid disease detection and monitoring
- 2 Reliable Stabilization of N-linked Polypeptide Native States With Enhanced Aromatic Sequons Located in Polypeptide Tight Turns
- 3 Small Molecules that Covalently Modify Transthyretin and Their Preparation
- 4 Methods for Identifying Cellular Modulators of Disaggregation Activity or Aggregation Activity in an Animal
- 5 Genistein Inhibition of transthyretin amyloidosis
- 6 Preparation of bisaryloxime ethers and hydrazones as Inhibitors of Transthyretin Amyloid Fibril Inhibitors
- 7 Preparation of Dibenzofuran-4,6-dicarboxylic acid Derivatives for Transthyretin Stabilization
- 8 Anti-amyloidogenic Agents