JEFFERY W. KELLY

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
Co-Chairman, Department of Molecular Medicine, Scripps, 2017-
Chairman, Molecular and Experimental Medicine, Scripps, 2008-2017
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-
Board of Directors, George E. Hewitt Foundation for Medical Research 2013-
Board of Directors, Amyloidosis Research Consortium 2015-
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-

Awards:

American Institute of Chemists Chemical Pioneer Award, 2017
Jacob and Louise 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)

Partial List of Academic Service:

The Celiac Sprue Research Foundation-Scientific Advisory Board Member ,2002-
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 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

41 of 100 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** Lawerence 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
Derrick Ong Graduate Student	Ph.D.	**Assistant Professor** School of Medicine National University of Singapore

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

Lars Plate Ph.D. **Assistant Professor**
Post-doctoral Fellow Department of Chemistry
Vanderbilt University

Current Funding

R37 DK046335 (Kelly) 08/01/11 – 07/31/16
NIH/NIDDK $331,900
Probing the Biochemical Mechanism of Amyloid Disease
This project is centered on understanding the chemistry and biology of transthyretin amyloid diseases using patient samples, organismal models, as well as in vitro studies.

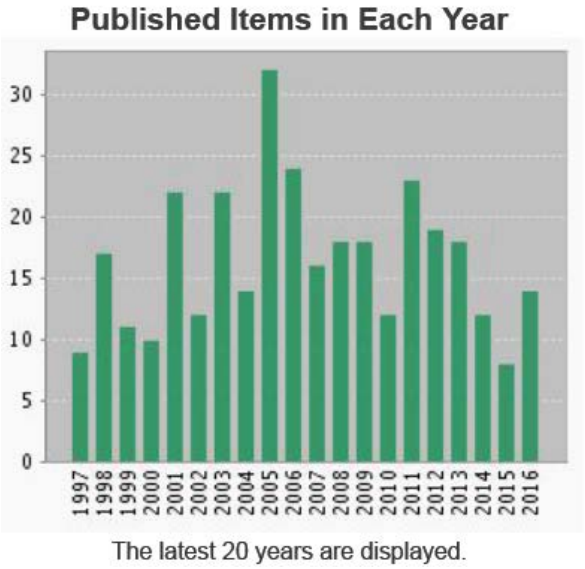
R01 GM051105 (Kelly) 05/01/12 – 02/29/16
NIH $180,000
Understanding β-sheet Structure in Aqueous Solution
This project is focused on understanding the kinetics and thermodynamics of β-sheet folding as it relates to the thermodynamic linkage of hydrogen bonding to the hydrophobic effect and also explores the chemistry and biology of the folding of glycosylated receptors.

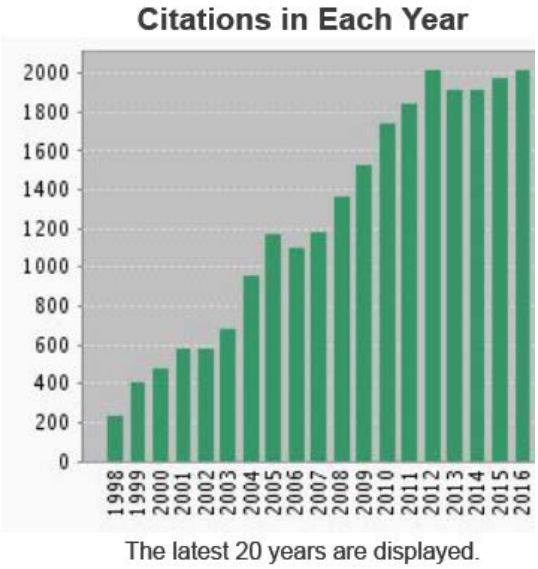
R01 AG046495 (Kelly) 04/01/13-03/31/18
NIH $202,950
Discovering Small Molecule Regulators of the Proteostasis Network
This project is focused on discovering small molecules that can selectively activate stress-responsive signaling pathways including the heat shock response (HSR) regulating proteostasis in the cytosol; the unfolded protein response (UPR) affecting proteostasis in the endoplasmic reticulum, the secretory pathway and the extracellular milieu; and the Daf-16/FOXO transcriptional program balancing metabolic and proteostatic control.

R01 DK106582 (Kelly)
NIH/NIDDK
Interplay of Intrinsic and Extrinsic Effects of N-glycans on Glycoproteostasis
In this proposal, we combine cell biology and biophysical approaches to study the factors that affect trafficking vs. degradation decisions by the glycoproteostasis network. We will examine how the initial N-glycosylation event by oligosaccharyl transferase influences downstream trafficking vs. degradation (i.e., quality control) decisions by the glycoproteostasis network. In addition, we will determine how the conformational properties of the N-glycoprotein determine the processing of N-glycans by glycoproteostasis network components.

Publication List–Jeffery W. Kelly:

ISI ***h-index* = 87**, *87 publications with 87 or more citations*
(> 70 papers with ≥ 100 citations, 25,500+ total citations)
Google Scholar ***h-index* > 100**





Data from ISI database as of March 14, 2017

---

330 Huang, Y-W.; Yang, H-I.; Wu Y-T.; Hsu, T-L.; Lin, T-W.; Kelly, J.W.; Wong, C-H. "Residues Comprising the Enhanced Aromatic Sequon Influence Protein N-glycosylation Efficiency" *J. Am. Chem. Soc.* **2016** xxx, yyyy-zzzz.

329 Maziar, A.; Powers, E.T.; Kelly, J.W. "Using Cooperatively Folded Peptides to Measure Interaction Energies and Conformational Propensities" *Acct. Chem. Res.* **2017** xx, yyy-zzz.

328 Schonhoft, J.D.; Monteiro, C.; Plate, L.; Eisele, Y.S.; Kelly, J.; Boland, D.; Parker, C.G.; Cravatt, B.F.; Berk, J.; Sekijima, Y.; Maurer, M.; Novias, M.; Coelho, T.; Powers, E.T.; Kelly, J.W. "Peptide Probes Detect Misfolded Transthyretin Oligomers in the Plasma of Early Stage Hereditary Amyloidosis Patients" *Science Trans. Med.* **2017** *in press*

327 Chen, K-C; Qu, S.; Chowdhury, S.; Noxon, I.C.; Schonhoft, J.D.; Plate, L.; Powers, E.T.; Kelly, J.W.; Lander, G.C.; Wiseman, R.L. "The ER HSP40 Co-chaperone ERdj3/DNAJB11 Assembles and Functions as a Native Tetramer" *EMBO J.* **2017** 36, 2296-2309.

326 Connelly, S.; Mortenson, D.E.; Choi, S.; Wilson, I.A.; Powers, E.T.; Kelly, J.W.; Johnson, S.M. “Semi-Quantitative Models for Identifying Potent and Selective Transthyretin Amyloidogenesis Inhibitors” *Biorg. Med. Chem. Lett.* **2017** 27 3441-3149.

325 Fernandes, L.; Moraes, N.; Sagrillo, F.; Magalhães, A.; de Moraes, M.; Romão, L.; Kelly, J.W.; Foguel, D; Grimster, N.; Palhano, F. “An ortho-Iminoquinone Compound Reacts with Lysine Inhibiting Aggregation while Remodeling Mature Amyloid Fibrils” *ACS Chemical Neuroscience* **2017** *in press*

324 Alvarez-Garcia, O.; Matsuzaki, T.; Olmer, M.; Plate, L.; Kelly, J.W.; Lotz, M.K. “REDD1 deficiency impairs autophagy and mitochondrial biogenesis in articular cartilage and increases severity of experimental osteoarthritis” Arthritis and Rheumatology **2017** 69, 1418-1428,

323 Lim, K.H.; Dasari, A. K.R.; Hung, I.F.; Gan, Z.; Kelly, J.W.; Wright, P.E. Wemmer, D.E. “Solid-State NMR Studies Reveal Native-like β-sheet Structures in Transthyretin amyloid” Biochemistry **2016** *55* 5272-5278.

322 Chen, J.J.; Genereux, J.C.; Suh, E.H.; Vartabedian, V.F.; Qu, S.; Dendle, M.T.; Kelly, J.W.; Wiseman, R.L. “Endoplasmic Reticulum Proteostasis Influences the Oligomeric State of an Amyloidogenic Protein Secreted from Mammalian Cells" *Cell Chem. Biol.* **2016** 23, 1282-1293.

321 Morgan, G. J.; Kelly, J.W. “The Kinetic Stability of a Full-length Antibody Light Chain Dimer Determines Whether Endoproteolysis can Release Amyloidogenic Variable Domains” *J. Mol. Biol.* **2016** 428, 4280-4297.

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.; Fearns, 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.; Poon, 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.; Fearns, 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.

298 Greiner, E.R.; Kelly, J.W.; Palhano, F.L. "Immunoprecipitation of Amyloid Fibrils by the use of an Antibody that Recognizes a Generic Epitope Common to Amyloid Fibrils" PLOS One **2014** 9(8), e105433 12pp.

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.; Hennegan, 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 Abeta 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.; Conceiçao, 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.

280 Shoulders, M.D.; Ryno, L.M.; Kelly, J.W.; Wiseman, R.L. "Broadly Applicable Methodology for the Rapid and Dosable Small-molecule Mediated Regulation of Transcription Factors in Human Cells" *J. Am. Chem. Soc.* **2013** 135, 8129-8132.

279 Talantova, M.; Sanz-Blasco, S.; Zhang, X.; Xia, P.; Akhtar, M.W.; Okamoto, S; Dziewczapolski, G.; Nakamura, T.; Cao, G; Pratt, A.E.; Kang, Y-J; Tu, S.; Molokanova, S.; McKercher, S.R.; Hires, S.A.; Sason, H.; Stouffer, D.G.; Buczynski, D.C.; Solomon, J.C.; Michael, S.; Powers, E.T.; Kelly, J.W.; Roberts, A.; Tong, G.; Fang-Newmeyer, T.; Parker, J.; Holland, E.A.; Zhang, D.; Nakanishi, N.; Chen, H.S.; Wolosker, H.; Parsons, L.H.; Ambasudhan, R.; Masliah, E.; Heinemann, S.F.; Pia-Crespo, J.C.; Lipton, S.A. "Aβ induces astrocytic glutamate release, extrasynaptic NMDA receptor activation, and synaptic loss" *Proc. Natl. Acad. Sci.* **2013** 110, E2518-2527, SI=10.

278 Palhano, F.L.; Lee, J.; Grimster, N.P.; Kelly, J.W. “Toward the Molecular Mechanism(s) by Which EGCG Remodels Mature Amyloid Fibrils” *J. Am. Chem. Soc.* **2013** 135, 7503-7510.

277 Shoulders, M.D.; Ryno, L.M.; Genereux, J.C.; Moresco, J.J.; Tu, P.G. Wu, C.; Yates, J.R.; Su, A.I.; Kelly, J.W.; Wiseman, R.L. “Stress-Independent Activation of XBP1s and/or ATF6 Reveals Three Functionally Diverse ER Proteostasis Environments” *Cell Reports* **2013** 3, 1279-1292.

276 Murray, A.N.; Palhano, F.L.; Bieschke, J.; Kelly, J.W. “The Challenges of amyloid disaggregation experiments performed in multi-well plates and Eppendorf tubes” *Protein Sci.* **2013** 22, 1531-1541.

275 Grimster, N.P.; Connelly, S.; Baranczak, A.; Dong, J.; Krasnova, L.B.; Sharpless, K.B.; Powers, E.T.; Wilson, I.A.; Kelly, J.W. “Aromatic Sulfonyl Fluorides Covalently Kineticlly Stabilize Transthyretin to Prevent Amyloidogenesis while Affording a Fluorescent Conjugate” *J. Am. Chem. Soc.* **2013** 135, 5656-5668.

274 Lim, Kwang Hun; Dyson, H. Jane; Kelly, J.W.; Wright, P.E. “Localized Structural Fluctuations Promote Amyloidogenic Conformations in Transthyretin” *J. Mol. Biol.* **2013** 425, 977-988.

273 Ong, D.S.T.; Wang, Y-J.; Tan, Y-L.; Yates, J.R.; Mu, T-W.; Kelly, J.W. “FKBP10 is an Endoplasmic Reticulum Degradation vs. Protein Folding Partitioning Factor” *Chem. And Biol.* **2013** 20, 403-415.

272 Azevedo, E.P.C; Guimaraes-Costa, A.B.;Torezani, G.S.; Braga, C.A.; Palhano, F.L.; Kelly, J.W.; Saraiva, E.M.; Foguel, D. “Amyloid Fibrils Trigger the Release of Neutrophil Extracellular Traps (NETs) Causing Fibril Fragmentation by NET-associated Elastase” *J. Biol. Chem.* **2012** 287, 37206-37218.

271 Fowler, D.M.; Kelly, J.W. “Functional Amyloidogenesis and Cytotoxicity–Insights into Biology and Pathology” *PLoS Biol.* **2012** 10, e1001459

270 Murray, A.; Kelly, J.W. “Hsp104 Gives Clients the Individual Attention They Need” *Cell* **2012** 151, 695-7.

269 Hulleman, J.D.; Brown, S.J.; Rosen, H.; Kelly, J.W. “A High-throughput Cell-based Gaussia Luciferase Reporter Assay for Identifying Modulators of Fibulin-3 Secretion” *J. Biomol. Screening.* **2012** 18, 647-658.

268 Araya, C.L.; Fowler, D.M.; Chen, W; Kelly, J.W.; Fields, S. “A Fundamental Protein Property, Thermodynamic Stability, Revealed Solely From Large-Scale Measurements of Protein Function” *Proc. Natl. Acad. Sci.* **2012** 109, 16858-16863.

267 Hulleman, J.D.; Balch, W.E.; Kelly, J.W. “Translational Attenuation Differentially Alters the Fate of Disease-associated Fibulin Proteins” *FASEB J.* **2012** 26, 4548-4560.

266 Bulawa, C.E.; Connelly, S.; DeVit, M.; Wang, L. Weigel, C.;Fleming, J. Packman, J.; Powers, E.T.; Wiseman, R.L.; Foss, T.R.; Wilson, I.A.; Kelly, J.W.; Labaudiniere, R. “Tafamidis, A Potent and Selective Transthyretin Kinetic Stabilizer That Inhibits the Amyloid Cascade” *Proc. Natl. Acad. Sci.* **2012** 109, 9629-9634.

265 Berk, J; Suhr, O.B.; Sekijima, Y.; Yamashita, X.; Heneghan, X.; Zeldenrust, S.; Ando, Y.; Ikeda, X.; Gorevic, P.; Merlini, P.; Kelly, J.W.; Skinner, M.; Bisbee, X.; Dyck, P.; Obici, X. “The Diflunisal Trial: Study Accrual and Drug Tolerance” *Amyloid* **2012** 19, 37-38.

264 Bett, C.; Joshi-Barr, S.; Lucero, M.; Trego, M.; Liberski, P.; Kelly, J.W.; Masliah, E.; Sigurdson, C.J. “Biochemical Properties of Highly Neuroinvasive Prion Strains” *PLoS Pathogens* **2012** 8, e1002522.

263 Coelho, T.; Maia, L.F.; Martins da Silva, A.; Cruz, M.W.; Planté-Bordeneuve, V.; Lozeron, P.; Suhr, O.B.; Campistol, J.M.; Conceiçao, I.; Schmidt, H.; Trigo, P. Kelly, J.W.; Labaudiniere, R.; Chan, J., Packman, J.; Wilson, A.; Grogan, D.R. “Tafamidis for transthyretin familial amyloid polyneuropathy: a randomized, controlled trial” *Neurology* **2012**, 79, 785-792.

262 Solomon, J.P.; Page, L.J.; Balch, W.E.; Kelly, J.W. “Gelsolin Amyloidosis: Genetics, Biochemistry, Pathology and Possible Strategies for Therapeutic Intervention” *Crit. Rev. Biochem. Mol. Biol.* **2012** 47, 282-296.

261 Price, J.L.; Culyba, E.K.; Chen, W.; Murray, A.; Hanson, S.R.; Wong, C.-H.; Powers, E.T.; Kelly, J.W. “N-glycosylation of the Enhanced Aromatic Sequon to Increase Glycoprotein Stability” *Peptide Sci.* **2012** 98, 195-211.

260 Johnson, S.M.; Connelly, S.; Fearns, C.; Powers, E.T.; Kelly, J.W. “The Transthyretin Amyloidoses: From Delineating the Molecular Mechanism of Aggregation Linked to Pathology to a Regulatory Agency Approved Drug” *J. Mol. Biol.* **2012** 421, 185-203.

259 Holtzman, D.M.; Goate, Alison; Kelly, J.W.; Sperling, R. “Mapping the Road Forward in Alzheimer’s Disease” *Science Trams. Med.* **2011**, 3, 114ps48. DOI:10.

258 Kelly, J.W. “Finally, a Light Chain Amyloidosis Transgenic Mouse Model” *Blood* **2011** 118, 6479-6480.

257 Azevedo, E,; Pereira, H.; Garratt, R.; Kelly, J.W.; Foguel, D; Palhano, F. “Dissecting the structure, thermodynamic stability and aggregation properties of the A25T transthyretin (A25T-TTR) variant involved in leptomeningeal

amyloidosis: identifying protein partners that co-aggregate during A25T-TTR fibrillogenesis in cerebrospinal fluid" *Biochemistry* **2011** 50, 11070-11083.

256 Orwig, S.D.; Tan, Y.L.; Grimster, N.P.; Yu, Z.; Powers, E.T.; Kelly, J.W.; Lieberman, R.L "Binding of 3,4,5,6-tetrahydroxyazepanes to the acid-β-glucosidase active site: implications for pharmacological chaperone design for Gaucher disease" *Biochemistry* **2011** 50, 10647-10657.

255 Hulleman, J.D.; Kaushal, S.; Balch, W.E.; Kelly, J.W. "Compromised EFEMP1 Secretion Associated with Macular Dystrophy Remedied by Proteostasis Network Alteration" *Mol. Biol. Cell* **2011** 22, 4765-4775.

254 Lindquist, S.L.; Kelly, J.W. "Chemical and Biological Approaches for Adapting Proteostasis to Ameliorate Protein Misfolding and Aggregation Diseases–Progress and Prognosis" *Cold Spring Harbor Perspect. Biol.* **2011**, 3(12) 307-340 (doi10.1101 / cshperspect.a004507).

253 Price, J.L.; Powers, E.T.; Kelly, J.W. "N-PEGylation of a Reverse Turn is Stabilizing in Multiple Sequence Contexts unlike N-GlcNAcylation" *ACS Chem. Biol.* **2011**, 6, 1188-1192.

252 Alhamadsheh, M.M.; Connelly, S.; Reixach, N.; Powers, E.T.; Pna, D.W.; Wilson, I.A.; Kelly, J.W.; Graef, I.A. "Potent Kinetic Stabilizers that Prevent Transthyretin-mediated Cardiomyocyte Proteotoxicity" *Science Translational Med.* **2011**, 97ra81.

251 Bourgault, S.; Choi, S-W.; Buxbaum, J.N.; Kelly, J.W.; Price, J.L.; Reixach, N. "Mechanisms of Transthyretin Cardiomyocyte Toxicity Inhibition by Resveratrol Analogs" *Biochem. Biophys. Res. Comm.* **2011**, 410, 707-713.

250 Price, J.L.; Powers, D.L.; Powers, E.T.; Kelly, J.W. "Glycosylation of the Enhanced Aromatic Sequon is Similarly Stabilizing in Three Distinct Reverse Turn Contexts" *Proc. Natl. Acad. Sci.* **2011**, 108, 14127-14132.

249 Reinhart, P.H.; Kelly, J.W. "Treating the Periphery to Ameliorate Neurodegenerative Diseases" *Cell.* **2011**, 145, 813-814.

248 Lee, J.; Culyba, E.K.; Powers, E.T.; Kelly, J.W. "Amyloid-β Fibrils form by Nucleated Conformational Conversion of Oligomers" *Nature Chem. Biol.* **2011**, 7, 602-609.

247 Ong, D.S.T.; Kelly, J.W. "Chemical and/or Biological Therapeutic Strategies to Ameliorate Protein Misfolding Diseases" *Curr. Op. Cell. Biol.* **2011**, 23, 231-238.

246 Wiseman, R.L.; Kelly, J.W. "Phosphatase Inhibition Delays Translational Recovery" *Science* **2011**, 332, 44-45.

245 Solomon, J.P.; Bourgault, S.; Powers, E.T.; Kelly, J.W. "Heparin Binds 8 kDa Gelsolin Cross-β-Sheet Oligomers and Accelerates Amyloidogenesis by Hastening Fibril Extension" *Biochemistry* **2011** 50, 2486-2498.

244 Bourgault, S.; Solomon, J.P.; Reixach, N.; Kelly, J.W. "Sulfated Glycosaminoglycans Accelerate Transthyretin Amyloidogenesis by Quaternary Structural Conversion" *Biochemistry* **2011** 50, 1001-1015.

243 Du, D.; Murray, A.; Cohen, E.; Kim, H-E; Simkovsky, R.; Dillin, A.; Kelly, J.W. "A Kinetic Aggregation Assay Enabling Selective and Sensitive Aβ Amyloid Quantification in Cells and Tissues" *Biochemistry* **2011** 50, 1607-1617.

242 Choi, S.; Kelly, J.W. "A Competition Assay to Identify Amyloidogenesis Inhibitors by Monitoring the Fluorescence Emitted by the Covalent Attachment of a Stilbene Derivative to Transthyretin" *Bioorg. Med. Chem.* **2011** 19, 1505-1514.

241 Culyba, E.K.; Price, J.L.; Hanson, S.R.; Dhar, A,; Wong, C-H.; Gruebele, M.; Powers, E.T.; Kelly, J.W. "Protein Native State Stabilization by Placing Aromatic Side Chains in N-Glycosylated Reverse Turns" *Science* **2011**, 331, 571-575.

240 Choi, S.; Ong, D.S.T.; Kelly, J.W. "A Stilbene that Binds Selectively to Transthyretin in Cells and Remains Dark Until it Undergoes a Chemoselective Reaction to Create a Bright Blue Fluorescent Conjugate" *J. Am. Chem. Soc.* **2010** 132, 16043-16051.

239 Price, J.L.; Shental-Bechor, D.; Dhar, A.; Turner, M.J.; Powers, E.T.; Gruebele, M.; Levy, Y.; Kelly, J.W. "Context-Dependent Effects of Asparagine Glycosylation on Pin WW Folding Kinetics and Thermodynamics" *J. Am. Chem. Soc.* **2010**, 132, 15359-15367.

238 Trivella, D.B.B.; Bleicher, L.; Palmieri, L.; Wiggers, H.J.; Montanari, C.A.; Kelly, J.W.; Lima, L.M.T.R.; Foguel, D.; Polikarpov, I. "Confromational Differences Between the Wild Type and V30M Mutant Transthyretin Modulate its Binding to Genistein: Implications for Tetramer Stability and Ligand Binding *J. Struct. Biol.* **2010**, 170, 522-531.

237 Murray, A.N.; Solomon, J.P.; Balch, W.E.; Kelly, J.W. "Discovery and Characterization of a Mammalian Amyloid Disaggregation Activity" *Protein Sci.*, **2010**, 19, 836-840.

236 Ong, D. S. T.; Mu, T-W; Palmer, A.E.; Kelly, J.W. "Endoplasmic Reticulum Ca^{2+} Increases Enhance Mutant Glucocerebrosidase Proteostasis" *Nature Chem. Biol.* **2010**, 6, 424-432.

235 Koob, A.O.; Ubhi, K.; Paulsson, J.F.; Kelly, J.W.; Rockenstein, E.; Mante, M.; Adame, A.; Masliah, E. "Lovastatin ameliorates α-synuclein accumulation and

oxidation in transgenic mouse models of α-synucleinopathies" Exp. Neurol. **2010**, 221, 267-274.

234 Cohen, E.; Du, D.; Kapernick, E.A.; Joyce, D.; Kelly, J.W.; Dillin, A. "Temporal Requirements of Insulin/IGF-1 Signaling for Proteotoxicity Protection" *Aging Cell* **2010**, 9, 126-134.

233 Wiseman, R.L.; Kelly, J.W. "Evolving Protein Stability Through Genetic Selection" *Mol. Cell.* **2009**, 36 730-731.

232 Connelly, S.; Choi, S.; Johnson, S. M.; Kelly, J.W.; Wilson, I.A. "Structure-Based Design of Kinetic Stabilizers That Ameliorate the Transthyretin Amyloidoses" *Curr. Op. Struct. Biol.* **2010**, 20, 1-9.

231 Choi, S.; Reixach, N.; Connelly, S.; Johnson, S.M.; Wilson, I.A.; Kelly, J.W. "A Substructure Combination Strategy to Create Potent and Selective Transthyretin Kinetic Stabilizers that Prevent Amyloidogenesis and Cytotoxicity" *J. Am. Chem. Soc.* **2010**, 132, 1359-1370.

230 Choi, S.; Connelly, S.; Reixach, N.; Wilson, I.A.; Kelly, J.W. "Highly Chemoselective Small Molecules That Covalently Modify one Lys in a Non-enzyme Protein in Plasma" *Nature Chem. Biol.* **2010**, 6, 133-139.

229 Watt, B.; van Niel, G.; Fowler, D.M.; Hurbain, I.; Luk, K.C.; Stayrook, S.E.; Lemmon, M.A.; Raposo, G.; Shorter, J.; Kelly, J.W.; Marks, M.S. "N-Terminal Domains Elicit Formation of Functional Pmel17 Amyloid Fibrils" *J. Biol. Chem.* **2009**, 284, 35543-35555.

228 Cohen, E.; Cohen, T-B; Paulsson, J.F.; Du, D.; Estepa, G.; Pham, H.M.; Kelly, J.W.; Masliah, E.; Dillin, A. "Reduced IGF-1 Signaling Delays Alzheimer's-like Disease Progression in Mice" *Cell* **2009** 137, 1157-1169.

227 Solomon, J.P.; Yonemoto, I.T.; Murray, A.N.; Price, J.L.; Powers, E.T.; Kelly, J.W. "Plasma Gelsolin 5 and 8 kDa Fragments Form Amyloid by a Nucleated Polymerization Mechanism, while the 68 kDa Fragment is Not Amyloidogenic" *Biochemistry* **2009**, 48, 11370-11380.

226 Naik, S.; Haque, I.; Degner, N.; Kornlayev, B.; Bomhoff, G.; Hodges, J.; Khorassani, A-A.; Katayama, H.; Morris, J.; Kelly, J.W.; Seed, J.; Fisher; M.T. "Identifying Protein Stabilizing Ligands using GroEL" *Biopolymers* **2010** 93, 237-251.

225 Hutt, D.M.; Herman, D.M.; Rodriques, A.P.C.; Noel, S.; Pilewski, J.M.; Matteson, J.; Hoch, B.; Kellner, W.; Kelly, J.W.; Richardson, J.; Thomas, P.J.; Matsumura, Y.; Skach, W.R.; Gentzsh, M.; Riordan, J.R.; Sorscher, E.J.; Okiyonad, T.; Lukacs, G.L.; Frizzell, R.A.; Manning, G.; Gottesfeld, J.M.; Balch, W.E. "Reduced Histone

Deacetylase 7 Activity Restores Function to Misfolded CFTR in Cystic Fibrosis" *Nature Chem. Biol.* **2010** 6, 25-31.

224 Bieschke, J.; Cohen, E.; Murray, A.; Dillin, A.; Kelly, J.W. "A Kinetic Assessment of the *C.elegans* Amyloid Disaggregation Activity Enables Uncoupling of Disassembly and Proteolysis" *Protein Sci.* **2009** 18, 2231-2241.

223 Usui, K.; Hulleman, J.D.; Paulsson, J.F.; Siegel, S.J.; Powers, E.T.; Kelly, J.W. "Site-specific modification of Alzheimer's peptides by cholesterol oxidation products enhance aggregation energetics and neurotoxicity" *Proc. Natl. Acad. Sci.* **2009**, 106, 18563-18568.

222 Yonemoto, I.T.; Wood, M.R.; Balch, W.E.; Kelly, J.W. "A General Strategy for the Bacterial Expression of Amyloidogenic Peptides using BCL-XL-1/2 Fusions" *Protein Sci.* **2009** 18, 1978-1986.

221 Fuller, A.A.; Du, D.; Liu, F.; Davoren, J.E.; Kroon, G.; Case, D.A.; Dyson, H.J.; Powers, E.T.; Wipf, P.; Gruebele, M.; Kelly, J.W. "Evaluating β-turn Mimics as β-sheet Folding Nucleators" *Proc. Natl. Acad. Sci.* **2009** 106, 11067-11072.

220 Jager, M; Dendle, M.; Kelly, J.W. "Sequence Determinants of Thermodynamic Stability in a WW domain–an all β-sheet protein" *Protein Sci.* **2009** 18, 1806-1813.

219 Page, L.J.; Suk, J-Y.; Bazhenova, L.; Fleming, S.M.; Wood, M.; Guo, L.T.; M Misizin, A.P.; Kisilevsky, R.; Shelton, D.G.; Balch, W.E.; Kelly, J.W. "Secretion of Amyloidogenic Gelsolin Progressively Compromises Protein Homeostasis Leading to the Intracellular Aggregation of Proteins" *Proc. Natl. Acad. Sci.* **2009** 106, 11125-11130.

218 Bunagan, M.R.; Gao, J.; Kelly, J.W.; Gai, F. "Probing the Folding Transition State Structure of the Villin Headpiece Subdomain via Sidechain and Backbone Mutagenesis" *J. Am. Chem. Soc.* **2009** 131, 7470-7476.

217 Gao, J.; Bosco, D.A.; Powers, E.T.; Kelly, J.W. "Localized Thermodynamic Coupling between Hydrogen Bonding and Microenvironment Polarity Significantly Stabilizes Proteins" *Nature Struct. Mol. Biol.* **2009** 16, 684-690.

216 Fowler, D.M.; Kelly, J.W. "Aggregating Knowledge about Prions and Amyloid" *Cell* **2009** 137, 20-22.

215 Johnson, S.M.; Wiseman, R.L.; Reixach, N; Paulsson, J.; Choi, S.; Powers, E.T.; Buxbaum, J.N.; Kelly, J.W. "Understanding and Ameliorating the Transthyretin Amyloidoses" in "Protein Misfolding Diseases: Current and Emerging Principles", **2009** Wiley Blackwell-I also co-edited this book with Dobson and Ramirez-Alvardo who served as editor-in-chief.

214 Hanson, S.R.; Culyba, E.K.; Hsu, T-L.; Wong, C-H. Kelly, J.W,; Powers, E.T. "The Core Trisaccharide of an N-linked Glycoprotein Intrinsically Accelerates Folding and Enhances Stability" *Proc. Natl. Acad. Sci.* **2009** 106, 3131-3136.

213 Johnson, S.W.; Connelly, S.; Wilson, I.A.; Kelly, J.W. "Toward Optimization of the Second Aryl Substructure Common to Transthyretin Amyloidogenesis Inhibitors Using Biochemical and Structural Studies" *J. Med. Chem.* **2009** 52, 1115-1125.

212 Powers, E.T.; Morimoto, R.I.; Dillin, A.; Kelly, J.W.; Balch, W.E. "Biological and Chemical Approaches to Diseases of Proteostasis Deficiency" *Ann. Rev. Biochem.* **2009** 78, 959-91.

211 Sekijima, Y.; Kelly, J.W.; Ikeda, S–I "Pathogenesis of and therapeutic strategies to ameliorate the transthyretin amyloidoses" *Curr. Pharm. Design* **2008** 30, 3219-3230.

210 Jager, M.; Deechongkit, S.; Koepf, E.K.;Nguyen, H.;Gao, J.; Powers, E.T.; Gruebele, M.; Kelly, J.W. "Understanding the mechanism of beta-sheet folding from a chemical and biological perspective" *Biopolymers* **2008** 90, 751-758.

209 Johnson, S.; Connelly, S.; Wilson, I.; Kelly, J.W. "Toward Optimization of the Linker Substructure Common to Transthyretin Amyloidogenesis Inhibitors Using Biochemical and Structural Studies" *J. Med. Chem.* **2008** 51, 6348-6358.

208 Palaninathan, S.K.; Nilofar, N.M.M.; Snee, W.C.; Kelly, J.W.; Sacchettini, J.C. "Structural Insight Into the pH-Induced Conformational Changes Within the Native Human Transthyretin Tetramer" *J. Mol. Biol.* **2008** 382, 1157-1167.

207 Yonemoto, I.T.; Kroon, G.J.A.; Dyson, H.J.; Balch, W.E.; Kelly, J.W. "Amylin Proprotein Processing Generates Progressively More Amyloidogenic Peptides that Initially Sample the Helical State *Biochemistry* **2008** 47, 9900-9910.

206 Mu, T-W.; Ong, D.S.T.; Wang, Y-J; Balch, W. E.; Yates, J.R.; Segatori, L.; Kelly, J.W. "Chemical and Biological Approaches Synergize to Ameliorate Protein-Folding Diseases" *Cell* **2008** 134, 769-781.

205 Hurshman Babbes, A.R.; Powers, E.T.; Kelly, J.W. "Quantification of the Thermodynamically Linked Quaternary and Tertiary Structural Stabilities of Transthyretin and its Disease-Associated Variants–The Relationship between Stability and Amyloidosis" *Biochemistry* **2008** 47, 6969-6984.

204 Gao, J; Kelly, J.W. "Towards Quantification of Protein Backbone-Backbone Hydrogen Bonding Energies: An Energetic Analysis of an Amide-to-Ester Mutation in an α-Helix within a Protein" *Protein Sci.* **2008** 17, 1096-1101.

203 Liu, F.; Du, D.; Fuller, A. A.; Davoren, J. E.; Wipf, P.; Kelly, J.W.; Gruebele, M.

"An experimental survey of the transition between two-state and downhill protein folding scenarios" *Proc. Natl. Acad. Sci.* **2008** 105, 2369-74.

202 Mu, T-W.; Fowler, D. M.; Kelly, J. W. "Partial Restoration of Mutant Enzyme Homeostasis in Three Distinct Lysosomal Storage Disease Cell Lines by Altering Calcium Homeostasis" *PloS Biol.* **2008** 6, 253-265, *e26.*

201 Balch, W.E.; Morimoto, R.I.; Dillin, A.; Kelly, J.W. "Adapting Proteostasis For Disease Intervention" *Science* **2008** 319, 916-919.

200 Reixach, N.; Foss, T.R.; Santelli, E.; Pascual, J.; Kelly, J.W. "Human-Murine Transthyretin Heterotetramers are Kinetically Stable and Non-Amyloidogenic: A challenge For The Generation of Murine Models of Human Amyloidosis" *J. Biol. Chem.* **2008** 283, 2098-2107.

199 Bieschke, J.; Fu, J.; Siegel, S.J; Kelly, J.W. Alzheimer's Aβ Peptides Containing an Isostructural Backbone Mutation Afford Distinct Aggregate Morphologies but Analogous Cytotoxicity–Is there a Common Low–Abundance Toxic Structure(s)? *Biochemistry* **2008** 47, 50-59.

198 Johnson, S.M.; Connelly, S.; Wilson, I.A.; Kelly, J.W. "Biochemical and structural evaluation of highly selective 2-arylbenzoxazole-based transthyretin amyloidogenesis inhibitors" *J. Med. Chem.* **2008** 51, 260-270.

197 Munch, J.; Rucker, E.; Standker, L.; Adermann, K.; Goffinet, C.; Schindler, M.; Wildum, S.; Chinnadurai, R.; Rajan, D.; Fowler, D.; Kelly, J.W.; Keppler, O.; Forssmann, W-G.; Kirchoff, F. "Semen-derived Amyloidogenic Prostatic Acidic Phosphatase Fragments Dramatically Enhance HIV infection" *Cell* **2007** 131, 1059-1071.

196 Wiseman, R.L.; Powers, E.T.; Buxbaum, J.N.; Kelly, J.W.; Balch, W.E. "An Adaptable Standard For Protein Export from the Endoplasmic Reticulum" *Cell* **2007** 131, 809-821.

195 Dillin, A.; Kelly, J.W. "The Yin-Yang of Sirtuins" *Science* **2007** 317, 461-462.

194 Yu, Z; Sawkar, A.R.; Kelly, J.W. "Pharmacologic Chaperoning as a Strategy to Treat Gaucher Disease" *FEBS Lett.* **2007** 274, 4944-4950.

193 Wiseman, R.L.; Koulov, A.; Powers, E.T.; Kelly, J.W.; Balch, W.E. "Protein Energetics and Maturation of the Early Secretory Pathway" *Curr. Op. Cell Biol.* **2007** 19, 359-367.

192 Jager, M.; Dendle, M.; Kelly, J.W. "A Cross-strand Trp-Trp Pair Stabilizes a WW Domain at the Expense of Function" *Protein Sci.* **2007** 16, 2306-2313.

191 Stewart, C.R.; Wilson, L.M.; Zhang, Q.; Pham, C.L.L.; Waddington, L.J.; Staples, M.K.; Stapleton, D. Kelly, J.W., Howlett, G.J. “Oxidized Cholesterol Metabolites found in human Atherosclerotic Lesions Promote Apolipoprotein C-II Amyloid Fibril Formation” *Biochemistry* **2007** 46, 5552-5561.

190 Fowler, D.M.; Koulov, A.V.; Balch, W.E.; Kelly, J.W. “Functional Amyloid–From Bacteria to Humans *TIBS* **2007** 32, 217-224.

189 Jäger, M.; Nguyen, H.; Dendle, M.; Gruebele, M.; Kelly, J.W. “Influence of hPin1 WW N-terminal domain boundaries on function, protein stability and folding” *Protein Science* **2007** 16, 1495-1501.

188 Kelly, J.W. “Compromised Cellular Folding Fidelity Results in Numerous Clinically Important Diseases” *Nature* **2007** 446, 112-113.

187 Siegel, S.J.; Bieschke, J.; Powers, E.T.; Kelly, J.W. “The Oxidative Stress Metabolite 4-Hydroxynonenal promotes Alzheimer Protofibril Formation” *Biochemistry* **2007** 46, 1503-1510.

186 Yu, Zhanqian; Sawkar, A.R.; Whalen, L.J.; Wong, C-H; Kelly, J.W. “Isofagamine and 2,5-Anhydro-2,5-Imino-D-Glucitol-Based Glucocerebrosidase Pharmacological chaperones for Gaucher Disease Intervention” *J. Med. Chem.* **2007**, 50, 94-100.

185 Wang, X.; Venable, J.; LaPointe, P.; Hutt, D.M.; Koulov, A.V.; Coppinger, J.; Gurkan, C.; Kellner, W.; Matteson, J.; Plutner, H.; Riordan, J.R.; Kelly, J.W.; Yates, J.R.; Balch, W.E. “Hsp90 Cochaperone Aha1 Downregulation Rescues Misfolding of CFTR in Cystic Fibrosis” *Cell* **2006** 127, 803-815.

184 Fu, Y.; Bieschke, J.; Gao, J.; Dendle, M.A.; Kelly, J.W. “Amide-to-E-Olefin versus Amide-to-Ester Backbone H-bond Perturbations: Evaluating the O-O Repulsion For Extracting H-Bond Energies” *J. Am. Chem. Soc.* **2006** 128, 15948-15949.

183 Zhang, Y.; Kim, Y.; Genoud, N.; Gao, G.; Kelly, J.W.; Platt, S.; Gill, G.; Dixon, J.; Noel, J.P. “Determinants for Dephosphorylation of the RNA Polymerase-II C-Terminal Domain by Scp-1” *Molecular Cell*, **2006**, 24, 759-770.

182 Tojo, K.; Sekijima, Y.; Kelly, J.W.; Ikeda, S-I “Diflunisal stabilizes familial-amyloid polyneuropathy-associated transthyretin variant tetramers in serum against dissociation required for amyloidogenesis” *Neuroscience Research* **2006** 56, 441-449.

181 Reixach, N.; Adamanski-Werner, S.L.; Kelly, J.W.; Koziol, J.; Buxbaum, J.N. “Cell-based screening of inhibitors of transthyretin aggregation” *Biochem. Biophys. Res. Comm.* **2006** 348, 889-897.

180 Cordeiro, Y.; Kraineva, J.; Suarez M– C.; Tempesta, A-G.; Kelly, J.W.; Silva, J.L.; Winter, R.; Foguel, D. “Fourier transform infrared spectroscopy provides a fingerprint for the tetramer and for aggregates of transthyretin” *Biophys. J.* **2006** 91, 957-967.

179 Kelly, J.W. “Proteins downhill all the way” *Nature* **2006** 242, 255-256.

178 Bieschke, J.; Zhang, Q.; Bosco, D.A.; Lerner, R.A.; Powers, E.T.; Wentworth, P.; Kelly. J.W.; “Small Molecule Oxidation Products Trigger Disease-Associated Protein Misfolding” *Acct. Chem. Res.* **2006** 39, 611-619.

177 Kelly, J.W.; Balch, W.E. “The Integration of Cell and Chemical Biology in Protein Folding” *Nat. Chem. Biol.* **2006** 2, 224-227.

176 Jager, M.; Zhang, Y.; Bieschke, J.; Nguyen, H.; Dendle, M.; Bowman, M.E.; Noel, J.P.; Gruebele, M.; Kelly, J.W. “The Structure–Function–Folding Relationship in a WW Domain *Proc. Natl. Acad. Sci*. **2006** 103, 10648-10653.

175 Sawkar, A.R.; Schmitz, M.; Zimmer, K-P; Reczek, D.; Edmunds, T. Balch, W.E.; Kelly, J.W. “Chemical chaperones and permissive temperatures alter localization of Gaucher disease-associated glucocerebrosidase variants” *ACS Chemical Biology* **2006**, 1, 235-251.

174 Sekijima, Y.,;Dendle, M.; Kelly, J.W. " Orally Administered Diflunisal Stabilizes Transthyretin Against Denaturation–A New Therapeutic Strategy for Amyloidosis" *Amyloid* **2006** 13, 236-249.

173 Cohen, E.; Bieschke, J.; Perciavalle, R.; Kelly, J.W.; Dillin, A. “Opposing activities protect against age onset proteotoxicity” *Science* **2006**, 313 1604-1610.

172 Bosco, D.; Fowler, D.M.; Zhang, Q.; Nieva, J.; Powers, E.T.; Wentworth, P.; Lerner, R.A.; Kelly, J.W. “Elevated levels of Oxidized Cholesterol Metabolites in Lewy Body Disease Brains Accelerate Alpha-synuclein Fibrilization” *Nature Chem. Biol.* **2006**, 2, 249-253.

171 Songpon Deechongkit, Houbi Nguyen, Marcus Jager, Evan T. Powers, Martin Gruebele, Jeffery W. Kelly ”β-Sheet Folding Mechanisms From Perturbation Energetics” *Curr. Op. Struct. Biol.* **2006** 16, 94-101.

170 Suk; J-Y.; Zhang, F.; Balch, W.E.; Linhardt, R.; Kelly, J.W. “Heparin Accelerates Gelsolin Amyloidogenesis” *Biochemistry* **2006** 45, 2234-2242.

169 Soergjerd, K.; Ghafouri, B.; Jonsson, B-H.; Kelly, J.W.; Blond, S.Y.; Hammarstrom, P. “Retention of Misfolded Mutant Transthyretin by the Chaperone BiP/GRP78 Mitigates Amyloidogenesis” *J. Mol. Biol.* **2006** 356, 469-482.

168 Sekijima, Y.; Dendle, M.T.; Wiseman, R.L.; White, J.T.; D'Haeze, W.; Kelly, J.W. "R104H may suppress transthyretin amyloidogenesis by thermodynamic stabilization, but not by the kinetic mechanism characterizing T119M interallelic trans-suppression *Amyloid* **2006** 13, 57-66.

167 Fowler, D.M.; Koulov, A.V.; Alory-Jost, C.; Marks, M.S.; Balch, W.E; Kelly, J.W. "Functional Amyloid Formation Within Mammalian Tissue " *PLoS Biology* **2006**, 4, 100-107.

166 Page, L.J.; Suk, J-Y.; Lim, L-J.; Kelly, J.W.; Balch, W.E. "Metalloendoprotease Cleavage Triggers Gelsolin Amyloidogenesis" *EMBO J.* **2005** 24, 4124-4132.

165 Sawkar, A.R.; D'Haeze, W.D.; Kelly, J.W. "Therapeutic Strategies for Gaucher Disease, a Prototypical Lysosomal Storage Disorder" *Cellular and Molecular Life Sciences* **2006** 63, 1179-1192.

164 Sawkar, A.R.; Adamanski-Werner, S.L.; Cheng, W-H.; Piper, J.; Wong, C-H.; Postema, M.; Beutler, E.; Zimmer, K-P; Kelly, J.W. "Gaucher Disease Associated Glucocerebrosidases Show Mutation Dependent Chemical Chaperoning Profiles" *Chemistry and Biology* **2005** 12, 1235-1244.

163 Wiseman, R.L; Powers, E.T.; Kelly, J.W. "Partitioning Conformational Intermediates Between Competing Refolding and Aggregation Pathways: Insights into Transthyretin Amyloid Disease" *Biochemistry* **2005** 44, 16612-16623.

162 Foss, T.R.; Wiseman, R.L.; Kelly, J.W. "The Pathway by Which the Tetrameric Protein Transthyretin Dissociates" *Biochemistry* **2005** 44, 15525-15533.

161 Fu, Y.; Bieschke, J.; Kelly, J.W. "An E-olefin Dipeptide Isostere Synthesis Enables Backbone Hydrogen Bond Perturbation: Probing the Requirements for Alzheimer's Amyloidogenesis" *J. Am. Chem. Soc.* **2005** 127, 15366-15367.

160 Kelly, J.W. "Form and Function Instructions" *Nature* **2005** 437, 486-487.

159 Nguyen, H.; Jager, M.; Kelly, J.W.; Gruebele, M. "Engineering a β-Sheet Protein Towards the Folding Speed Limit" *J. Phys. Chem. B, Biophysical Chemistry* **2005** 109, 15182-15186.

158 Johnson, S.; Green, N.; Adamanski-Werner, S.; Kelly, J.W. "Native State Kinetic Stabilization as a Strategy to Ameliorate Protein Misfolding Diseases: A focus on the Transthyretin Amyloidoses" *Acct. Chem. Res.* **2005** 38, 911-921.

157 Powers, E.T; Deechongkit, S; Kelly, J.W. "Backbone-Backbone H-Bonds Make Context Dependent Contributions to Protein Folding Kinetics and Thermodynamics: Lessons from Amide to Ester Mutations" *Adv. Pro. Chem.* **2006** 72, 39-78.

156 Green, N.; Foss, T. Kelly, J.W. “Genistein, A Natural Product from Soy, is a Potent Inhibitor of Transthyretin Amyloidogenesis” *Proc. Natl. Acad. Sci.* **2005** 102, 14545-14550.

155 Frankenfield, K.N.; Powers, E.T.; Kelly, J.W. “Influence of the N-terminal Domain on the Aggregation Properties of the Prion Protein” *Protein Sci.* **2005** 14, 2154-2166.

154 Wiseman, R.L.; Green, N.S.; Kelly, J.W. “Kinetic Stabilization of an Oligomeric Protein Under Physiological Conditions Demonstrated by Lack of Subunit Exchange–Implications for Transthyretin Amyloidogenesis”*Biochemistry* **2005** 44, 9265-9274.

153 Premkumar, L; Sawkar, A.R. ;Boldin-Adamsky, S Toker, L.; Silman, I; Kelly, J.W; Futerman, A.H.; Sussman, J.L. “X-ray structure of Human Acid-β-glucosidase Covalently-Bound to Conduritol-β-Epoxide: Implications for Gaucher disease” *J. Biol. Chem.* **2005** 280, 23815-23819.

152 Zhang, Q.; Kelly, J.W. “ Cys-10 Mixed Disulfide Modifications Exacerbate Transthyretin Familial Variant Amyloidogenicity: a Likely Explanation for Variable Clinical Expression of Amyloidosis and the Lack of Pathology in C10S/V30M Mice?” *Biochemistry* **2005** 44, 9079-9085.

151 Deechongkit, S., Powers, E.T., You, S-L., Kelly, J.W. “Controlling the Morphology of Cross-β-Sheet Assemblies by Rational Design” *J. Am. Chem. Soc.* **2005** 127, 8562-8570.

150 Bieschke, J.; Zhang, Q.; Powers, E.; Lerner, R.A.; Kelly, J.W. “Oxidative Metabolites Accelerate Alzheimer’s Amyloidogenesis by a Two Step Mechanism, Obviating The Requirement for Nucleation”*Biochemistry* **2005** 44, 4977-4983.

149 You, S-L.; Kelly, J.W. "Total Synthesis of Didmolamides A & B” *Tetrahedron Lett*. **2005** 46, 2567-2570.

148 Petrassi, H.M.; Johnson, S.; Purkey, H.E.; Kelly, J.W. "Potent and Selective Structure-Based Dibenzofuran Inhibitors of Transthyretin Amyloidogenesis: Kinetic Stabilization of the Native State" *J. Am. Chem. Soc.* **2005**, 127, 6662-6671.

147 Wiseman, R.L.; Johnson, S.M.; Kelker, M.S.; Foss, T.; Wilson, I.A.; Kelly, J.W. “Kinetic Stabilization of an Oligomeric Protein By a Single Ligand Binding Event” *J. Am. Chem. Soc*. **2005** 127, 5540-5551.

146 Sekijima, Y., Wiseman, R.L., Matteson, J., Hammarström, P., Miller, S.R., Balch, W.E., Kelly, J.W. “Biological and Chemical Basis for Tissue Selective Amyloid Disease”*Cell* **2005** 121, 73-85.

145 Foss, T.R.; Kelker, M.S.; Wilson, I.A.; Kelly, J.W. “Kinetic Stabilization of the Native State by Protein Engineering: Implications for Inhibition of Transthyretin Amyloidosis” *J. Mol. Biol.* **2005** 347, 841-854.

144 Kelly, J.W. “Attacking Amyloid” *New. Engl. J. Med.* **2005** 352, 722-723. [16]

143 Razavi, H.; Powers, E.T.; Purkey, H.E.; Adamanski-Werner, S.L.; Chiang, K.P.; Dendle, M.T.A.; Kelly, J.W. “Design, Synthesis, and Evaluation of Oxazole Transthyretin Amyloidogenesis Inhibitors” *Bioorg. Med. Chem. Lett*. **2005** 15, 1075-1078.

142 Johnson, S.M., Petrassi, H.M., Palaninathan, S.K., Mohamedmohaideen, N.N., Purkey, H.E., Nichols, C., Chiang, K.P., Walkup, T., Sacchettini, J.C., Sharpless, K.B., Kelly, J.W. “Bisaryloxime Ethers as Potent Inhibitors of Transthyretin Amyloid Fibril Formation”*J. Med. Chem.* **2005** 48, 1576-1587.

141 You, S-L.; Kelly, J.W. "Total Synthesis Of Bistratamides F-I" *Tetrahedron.* **2005** 61, 241-249.

140 Deechongkit, S.; Dawson, P.E.; Kelly, J.W. “Towards Assessing the Position-Dependent Contributions of Backbone Hydrogen Bonding to β-Sheet Folding Thermodynamics Employing Amide-to-Ester Perturbations”*J. Am. Chem. Soc.* **2004** 126, 16762-16771.

139 Kelker, M.S.; Foss, T.R.; Petit, W.; Teyton, L.; Kelly, J.W.; Wuthrich, K.; Wilson, I.A. “Crystal Structure of Human Triggering Receptor Expressed on Myeloid Cells 1 (TREM-1) at 1.47 Å *J. Mol. Biol.* **2004**, 342, 1237-1248.

138 Page, L.; Huff, M.E.; Kelly, J.W., Balch, W.E. “Ca^{2+} binding protects against gelsolin amyloidogenesis” *Biochem. Biophys. Res. Comm.* **2004** 322, 1105-1110.

137 You, S-L.; Deechongkit, S.; Kelly, J.W. "Solid Phase Synthesis and Sterochemical Assignments of Tenuecyclamides A-D Employing Heterocyclic Amino Acids Derived From Commercially Available Fmoc α-Amino Acids” *Org. Lett.* **2004** 6, 2627-2630.

136 Deechongkit, S.; Nguyen, H.; Dawson, P.E.; Gruebele, M.; Kelly, J.W. “Context Dependent Contributions of Backbone H-Bonding to β-Sheet Folding Energetics” *Nature* **2004** 430, 101-105.

135 Purkey, H.E.; Palaninathan, S.K.; Kent, K.C.; Smith, C.; Safe, S.H.; Sacchettini, J.C.; Kelly, J.W. "Hydroxylated Polychlorinated Biphenyls Selectively Bind Transthyretrin In Blood and Inhibit Amyloidogenesis; Rationalizing Rodent PCB Toxicity" *Chemistry and Biology* **2004** 11, 1719-1728.

134 You, S-L.; Kelly, J.W. “Highly Efficient Enantiospecific Synthesis of Imidazoline-Containing Amino Acids Using Bis(triphenyl)oxodiphosphonium Trifluoromethane sulfonate *Org. Lett.* **2004** 6, 1681-1683.

133 Hurshman, A.; White, J.P., Powers, E.T., Kelly, J.W. "Transthyretin Aggregation under Partially Denaturing Conditions is a Downhill Polymerization" *Biochemistry* **2004** 43, 7365-7381.

132 Reixach, N; Deechongkit, S; Jiang, X. Kelly, J.W.; Buxbaum, J.N. “Tissue Damage in the Amyloidoses: Transthyretin Monomers and Nonnative Oligomers are the Major Cytotoxic Species in Tissue Culture” *Proc. Natl. Acad. Sci* **2004** 101, 2817-2822.

131 Zhang, Q; Powers, E.T.; Nieva, J; Huff, M.E.; Dendle, M.A.; Bieschke, J.; Glabe, C.G.; Eschenmoser, A.; Wentworth, P., Jr.; Lerner, R.A.; Kelly, J.W. "Metabolite Initiated Protein Misfolding May Trigger Alzheimer’s Disease" *Proc. Natl. Acad. Sci* **2004**, 101, 4752-4757.

130 Miller, S.R.; Sekijima, Y.; Kelly, J.W. “Native State Stabilization by NSAIDs Inhibits Transthyretin Amyloidogenesis from the Most Common Familial Disease Variants” *Laboratory Investigation* **2004** 84, 545-552.

129 Deechongkit, S.;You, S-L. Kelly, J.W. “Synthesis of All Nineteen Appropriately Protected Chiral α-Hydroxy Acid Equivalents of the α-Amino Acids for Boc Solid Phase Depsi-Peptide Synthesis” *Org. Lett.* **2004** 6, 497-500.

128 Huff, M.E.; Balch, W.E.; Kelly, J.W. “Pathological and Functional Amyloid Formation Orchestrated By The Secretory Pathway” *Curr. Opin. Struct. Biol.* **2003** 13, 674-682.

127 Kustedjo, K.; Deechongkit, S.; Kelly, J.W., Cravatt, B.F. "Recombinant Expression, Purification, and Comparative Characterization of Torsion A and its Torsion Dystonia-associated Variant ΔE-Torsin A" *Biochemistry* **2003** 42, 15333-15341.

126 Adamanski-Werner, S.L., Kumar, P.S., Sacchettini, J.C., Kelly, J.W. "Diflunisal Analogues Stabilize the Native State of Transthyretin. Potent Inhibition of Amyloidogenesis" *J. Med. Chem.* **2004** 47, 355-374.

125 You, S-L.; Kelly, J.W. “Highly Efficient Biomimetic Total Synthesis and Structural Verification of Bistratamides E and J from Lissoclinum Bistratum" *Chemistry-A European Journal* **2004** 10, 71-75.

124 You, S-L.; Kelly, J.W. "Total Synthesis of Dendroamide A: Oxazole and Thiazole Construction Using an Oxodiphosphonium Salt" *J. Org. Chem.* **2003** 68, 9506-9509.

123 DiDonato, M.; Craig, L.; Huff; M.E.; Thayer, M.M.; Cardosa, R.M.F.; Kassman, C.J.; Lo, T.P.; Bruns, C.K.; Powers, E.T.; Kelly, J.W.; Getzoff, E.D.; Tainer, J.A. "ALS Mutants of

Human Superoxide Dismutase Form Fibrous Aggregates Via Framework Destabilization" *J. Mol. Biol.* **2003** 332, 601-615.

122 Green, N.S.; Palaninathan, S.K.; Sacchettini, J.C.; Kelly, J.W. "Synthesis and Characterization of Potent Bivalent Amyloidosis Inhibitors that Bind Prior to Transthyretin Tetramerization" *J. Am. Chem. Soc.* **2003** 125, 13404-13414.

121 Foguel, D.; Suarez, M.C.; Ferrao-Gonzales, A.D.; Porto, T.C.R.; Palmieri, L.; Einsiedler, C.M.; Andrade, L.R.; Lashuel, H.A.; Lansbury, P.T.; Kelly, J.W.; Silva, J.L. "Dissociation of amyloid fibrils of α-synuclein and transthyretin by pressure reveals their reversible nature and the formation of water excluded cavities" *Proc. Natl. Acad. Sci.* (USA) **2003** 100, 9831-9836.

120 Huff, M.E., Page, L., Balch, W.E., Kelly, J.W. "Gelsolin Domain 2 Ca^{2+} Affinity Determines Susceptibility to Furin Proteolysis and Familial Amyloidosis of Finnish Type" *J. Mol. Biol.* **2003** 334, 119-127.

119 Calarese, D.A.; Scanlan, C.N.; Zwick, M.B.; Deechongkit, S.Mimura, Y.; Kunert, R.; Zhu, P.; Wormald, M.R.; Stanfield, R.L.; Roux, K.H.; Kelly, J.W.; Rudd, P.M.; Dwek, R.A.; Katinger, H.;Burton, D.R.; Wilson, I.A. "Antibody Domain Exchange is an Immunological Solution to Carbohydrate Cluster Recognition" *Science* **2003**, 300, 2065-2071.

118 Ferrao-Gonzales, A.D.; Palmieri, L.; Valory, M.; Silva, J. L.; Lashuel, H.; Kelly, J.W.; Foguel, D. "Hydration and Packing are Crucial to Amyloidogenesis as Revealed by Pressure Studies on Transthyretin Variants that either Protect or Worsen Amyloid Disease" *J. Mol. Biol.* **2003**, 328, 963-974.

117 Zhang, Q.; Kelly, J.W. "Cys 10 Mixed Disulfides Make Transthyretin More Amyloidogenic Under Mildly Acidic Conditions" *Biochemistry* **2003** 42, 8756-8761.

116 Kelly, J.W.; Balch, W.E. "Amyloid As A Natural Product" *J. Cell Biol*. **2003**, 161, 461-462.

115 Cohen, F.; Kelly, J.W. "Therapeutic Approaches to Protein-Misfolding Diseases" *Nature*, **2003**, 426, 905-909.

114 Lu, J.R.; Perumal, S.; Powers, E.T.; Kelly, J.W.; Webster, J.R.P.; Penfold, J. "Adsorption of β-Hairpin Peptides on the Surface of Water: A Neutron Refection Study" *J. Am. Chem. Soc.*, **2003**, 125, 3751-3757.

113 Razavi, H.; Palaninathan, S.K. Powers, E.T.; Wiseman, R.L.; Purkey, H.E.; Mohamadmohaideen, N.N.; Deechongkit, S.; Chiang, K.P.; Dendle, M.T.A.; Sacchettini, J.C.; Kelly, J.W. "Benzoxazoles as Transthyretin Amyloid Fibril Inhibitors: Synthesis, Evaluation and Mechanism of Action" *Angew. Chem. Int. Ed.* **2003**, 42, 2758-2761.

112 Hammarstrom, P.; Sekijima, Y.; White, J.P.; Wiseman, R.L.; Lim, A.; Costello, C.E.; Atland, K., Garzuly, F.; Budka, H.; Kelly, J.W. "D18G Transthyretin is Monomeric, Aggregation Prone and Not detectable in Plasma and Cerebral Spinal Fluid-A Perscription of CNS Amyloidosis ?" *Biochemistry* **2003**, 42, 6656-6663.

111 Nguyen, H.; Jager, M.; Moretto, A.; Gruebele, M.; Kelly, J.W. "Tuning the free–energy landscape of a WW domain by temperature, mutation and truncation" *Proc. Natl. Acad. Sci.* **2003** 100, 3948-3953.

110 Colfer, S., Kelly, J.W.; Powers E.T. "Factors Governing the Self-Assembly of a β-Hairpin Peptide at the Air-Water Interface" *Langmuir* **2003** 19, 1312-1318.

109 Sekijima, Y.; Hammarstrom, P.; Matsumura, M.; Shimizu, Y.; Iwata, M.; Tokuda, T.; Ikeda, S-I.; Kelly, J.W. "Energetic Characteristics of the New Transthyretin Variant A25T May Explain Its Atypical Central Nervous System Pathology " *Lab. Inv.* **2003**, 83, 409-418.

108 Hammarstrom, P.; Wiseman, R. L.; Powers, E.T.; Kelly, J.W. "Prevention of Transthyretin Amyloid Disease by Changing Protein Misfolding Energetics" *Science* **2003**, 299, 713-716.

107 Kelly, J.W. "Towards an Understanding of Amyloidogenesis" *Nature Struct. Biol*. **2002**, 9, 323-325.

106 Sawkar, A.R.; Cheng, W-C.; Beutler, E.: Wong, C.–H.: Balch, W.E.: Kelly, J.W. "Chemical Chaperones Increase the Cellular Activity of N370S β-glucosidase: A Therapeutic Strategy for Gaucher Disease " *Proc. Natl. Acad. Sci*., **2002**, 99, 15428-15433.

105 You, S-L.; Razavi, H.; Kelly, J.W. "A Biomimetic Synthesis of Thiazolines Using hexaphenyloxodiphosphonium trifluoromethanesulfonate Reagents" *Angew. Chem. Int. Ed.* **2003**, 42, 83-85.

104 Kaul, R.; Deechongkit, S.; Kelly, J.W. "Synthesis of a Negatively-Charged Dibenzofuran-based β-Turn Mimetic and its Incorporation into the WW Miniprotein-Enhanced Solubility Without a loss of Thermodynamic Stability" *J. Am. Chem. Soc.* **2002**, 124, 11900-11907.

103 Hammarström, P.; Jiang, X.; Hurshman, A.R.; Kelly, J.W. "Sequence–Dependent Denaturation Energetics: A Major Determinant in Amyloid Disease Diversity" *Proc. Natl. Acad. Sci.* **2002**, 99, 16427-16432.

102 Liu, K.; Kelly, J.W.; Wemmer, D.E., Native State Hydrogen Exchange Study of Suppressor and Pathogenic variants of Transthyretin" *J. Mol. Biol*. **2002**, 320, 821-832.

101 McCammon, M.G.; Scott, D.J.; Keetch, C.A.; Greene, L.H.; Purkey, H.E.; Petrassi, H.M.; Kelly, J.W.; Robinson, C.V. "Screening Transthyretin Amyloid Fibril Inhibitors. Characterisation of Novel MultiProtein, MultiLigand Complexes by Mass Spectrometry *Structure* **2002**, 10, 851-863.

100 Deechongkit, S.; Kelly, J.W. "The Effect of backbone Cyclization on the Thermodynamics of β-Sheet Unfolding: Stability Optimization of the PIN WW Domain" *J. Am. Chem. Soc.* **2002**, 124, 4980-4986.

99 Kowalski, J.A.; Liu, K.; Kelly, J.W. "NMR Structure of the Isolated Apo Pin1 WW Domain; Comparison to the X-ray crystal structures of Pin 1" *Biopolymers* **2002**, 63, 111-121.

98 Sacchettini, J.C. and Kelly, J.W. "Therapeutic Strategies for Human Amyloid Diseases" *Nature Reviews Drug Discovery* **2002**, 1, 267-275.

97 Oza, V.B.; Smith, C.; Raman, P.; Koepf, E.K.; Laushel, H.A.; Petrassi, H.M.; Chiang,K.P.; Powers, E.T.; K.P.; Sacchettini, J.; Kelly, J.W. "Synthesis, Structure and Activity of Diclofenac Analogues as Transthyretin Amyloid Fibril Formation Inhibitors" *J. Med. Chem.* **2002**, 45, 321-332.

96 Powers, E.T.; Yang, S.I.; Lieber, C.M.; Kelly, J.W. "Ordered Langmuir-Blodgett Films of of Amphiphilic Peptides Imaged by Atomic Force Microscopy" *Angew. Chem. Int. Ed.* **2002**, 41, 127-130.

95 Jiang, X.; Buxbaum, J.N.; Kelly, J.W. "The V122I Cardiomyopathy Variant of Transthyretin Increases the Velocity of Rate-Limiting Tetramer Dissociation Resulting in Accelerated Amyloidosis" *Proc. Natl. Acad. Sci.*, **2001**, 98, 14943-14948.

94 White, J.T.; Kelly, J.W. "Support For the MultiGenic Hypothesis of Amyloidosis: The Binding Stoichiometry of retinol Binding Protein, Vitamin A, and Thyroid Hormone influences Transthyretin Amyloidogenicity in vitro" *Proc. Natl. Acad. Sci*., **2001**, 98 13019-13024.

93 Chen, C.D.; Huff, M. E; Matteson, J.; Page, L.; Phillips, R.; Kelly, J.W.; Balch, W.E. "Furin Initiates Gelsolin Familial Amyloidosis in the Golgi Through a Defect in Ca^{2+} Stabilization" *EMBO J.* **2001**, 20, 6277-6287.

92 Hammarstrom, P.; Schneider, F.; Kelly, J.W. "Trans-Suppression of Misfolding In An Amyloid Disease" *Science* **2001**, 293, 2459-2461.

91 Jager, M.; Nguyen, H.; Crane, J.C.; Kelly, J.W.; Gruebele, M. "The Folding Mechanism of a β-Sheet: The WW Domain" *J. Mol. Biol*. **2001**, 311, 373-393.

90 Hammarstrom, P.; Jiang, X.; Deechongkit, S.; Kelly, J.W. " Anion Shielding of Electrostatic Repulsions in Transthyretin Modulates Stability and Amyloidosis: Insight into the Chaotrope Unfolding Dichotomy " *Biochemistry* **2001**, 40, 11453-11459.

89 Jiang, X,; Smith, C.S.; Petrassi, H.M.; Hammarstrom, P.O.; White, J.; Sacchettini, J.C. Kelly, J.W. " An Engineered Transthyretin Monomer That is Non-Amyloidogenic-Unless It Is Partially Denatured" *Biochemistry* **2001**, 40, 11442-11452.

88 Schneider, F.; Hammarstrom, P.; Kelly, J.W. "Transthyretin slowly exchanges subunits under physiological conditions: A convenient chromatographic method to study subunit exchange in oligomeric proteins" *Protein Science* **2001**, 10, 1606-1613.

87 Jiang, X.; Kowalski, J.; Kelly, J.W. "Increasing protein stability using a rational approach combining sequence homology and structural alignment: Stabilizing the WW domain" *Protein Science* **2001**, 10, 1454-1465.

86 Kaul, R.; Angeles, A.R.; Jager, M.; Powers, E.T.; Kelly, J.W. " Incorporating Beta-turns and a Turn Mimetic out of context in Loop 1 of the WW Domain Affords Cooperatively Folded Beta-Sheets" *J. Am. Chem. Soc.*, **2001**, 123, 5206-5212.

85 Purkey, H.E.; Dorrell, M.I.; Kelly, J.W. "Evaluating the Binding Selectivity of Transthyretin Amyloid Fibril Inhibitors in Blood Plasma" *Proc. Natl. Acad. Sci.*, **2001,** 98, 5566-5571.

84 Ratnaswamy, G.; Huff, M.E.; Su, A.I.; Rion, S.; Kelly, J.W " Destabilization of Ca^{+2}-free gelsolin may not be responsible for proteolysis in Familial Amyloidosis of Finnish Type." *Proc. Natl. Acad. Sci*. **2001,** 98(5), 2334-2339.

83 Petrassi, H.M.; Sharpless, K.B.; Kelly, J.W. "The Copper–Mediated Cross–Coupling of Phenylboronic Acids and N-Hydroxy Phthalimide at Room Temperature: Synthesis of Aryloxyamines" *Org. Lett.* **2001**, 3, 139-142.

82 Powers, E.T.; Kelly, J.W. "Medium Dependent Self-Assembly of an Amphiphilic Peptide: Direct Observation of peptide Phase Domains at the Air-Water Interface" *J. Am. Chem. Soc.*, **2001**, 123, 775-776.

81 Kelly, J.W. "Mechanisms of Amyloidogenesis" *Nature-Struct. Biol.*, **2000**, 7, 824-6.

80 Liu, K.; Cho, H.S.; Hoyt, D.W.; Nguyen, T.N.; Olds, P.; Kelly, J.W.; Wemmer, D.E. "Deuterium-Proton Exchange on the Native WT Transthyretin Tetramer Identifies the Stable Core of Individual Subunits and Indicates Mobility at the Subunit Interface" *J. Mol. Biol.*, **2000**, 303, 555-565.

79 Chitnumsub, P.; Schneider, J.P.; Tsang, K.; Bekele, H.; Labrenz, S.R.; Lashuel, H.A.; Kelly, J.W. "Synthesis of Peptides Incorporating Templates" *Houben-Weyl*, **2000**.

78 Raman, P., Razavi, H., Kelly, J.W. "Titanium(IV) Mediated Tandem Deprotection-Cyclodehydration of Protected Cysteine N-Amides:Biomimetic Syntheses of Thiazoline– and Thiazole–Containing heterocycles" *Organic Letters*., **2000**, 2, 3289-3292.

77 Liu, K., Cho, H.S., Lashuel, H.A., Kelly, J.W., Wemmer, D.E. "A glimpse of A Possible Amyloidogenic Intermediate of Transthyretin" *Nature-Strutural Biology*, **2000**, 7, 754-757.

76 Lashuel, H.A.; LaBrenz,S.R.; Woo, L.; Serpell, L.C.; Kelly, J.W. "Protofilaments, Filaments, Ribbons and Fibrils From Peptidomimetic Self-Assembly: Implications

for Amyloid fibril Formation and Materials Science" *J. Am. Chem. Soc.*, **2000**, 122, 5262-5277.

75 Crane, J.C., Koepf, E.K.,Kelly, J.W., Gruebele, M. "Mapping the transition State of the WW Domain β-Sheet *J. Mol. Biol.*, **2000**, 298, 283-292.

74 Petrassi, H.M.; Klabunde, T.; Sacchettini, J..; Kelly, J.W. "Structure-Based Design of N-Phenyl Phenoxazine Transthyretin Amyloid Fibril Inhibitors" *J. Am. Chem. Soc*., **2000**, 122, 2178-2192.

73 Klabunde, T.; Petrassi, H. M.; Oza, V.B.; Raman, P.; Kelly, J.W.; Sacchettini, J.C.; "Rational Design of Potent Human Transthyretin Amyloid Disease Inhibitors" *Nature Struct. Biol*., **2000**, 7, 312-321.

72 Koepf, E.K., Petrassi, H.M., Ratnaswamy, G.; Huff, M.E.; Sudol, M., Kelly, J.W. "Characterization of the Structure and Function of W to F WW Domain variants: Identification of a Natively Unfolded Protein that Folds Upon Ligand Binding" *Biochemistry*, **1999**, 38, 14338-14351.

71 Lashuel, H.; Wurth, C., Woo, L.; Kelly, J.W. "The Most Pathogenic Transthyretin Variant, L55P, Forms Amyloid Fibrils Under Acidic Conditions and Protofilaments Under Physiological Conditions" *Biochemistry*, **1999**, 38, 13560-13573.

70 Koo, E.H.; Lansbury, P.T, Jr.; Kelly, J.W. "Amyloid Diseases: Abnormal Protein Aggregation in Neurodegeneration" *Proc. Natl. Acad. Sci. USA*, **1999**, 96, 9989-9990.

69 Bekele, H. Fendler, J.H., Kelly, J. W. "Self-Assembling Peptidomimetic Monolayer Nucleates Oriented CdS Nanocrystals *J. Am. Chem. Soc.*, **1999**, 121,7266-7267.

68 Oza, V.B.; Petrassi, H.M.; Purkey, H.E.; Kelly, J.W. "Synthesis and Evaluation of Anthranilic Acid-Based Transthyretin Amyloid Fibril Inhibitors *Bioorg. Med. Chem. Lett.* **1999**, 9, 1-6.

67 Kelly, J.W.; Editor, Peptides and Peptidomimetics that Adopt Folded Structures, Symposium in Print #15, *Bioorg. Med. Chem.*, **1999**, 7.

66 Ratnaswamy, G.; Koepf, E.; Bekele, H.; Yin, H.; Kelly, J. W. "The Amyloidogenicity of Gelsolin is Controlled by Proteolysis and pH" *Chemistry and Biology*, **1999**, 6, 293-304.

65 Baures, P.W.; Oza, V.H.; Peterson, S.A.; Kelly, J.W. "Synthesis and Evaluation of Inhibitors of Transthyretin Amyloid Fibril Formation Based on the Non-Steroidal Anti-Inflammatory Drug, Flufenamic Acid" *Biorg. Med. Chem.* **1999**, 7, 1339-1347.

64 Chitnumsub, P.; Fiori, W.R.;Lashuel, H.; Diaz, H.; Kelly, J.W. "The nucleation of monomeric Parallel β-sheet–like structures and their self-assembly in aqueous solution" *Bioorg. Med. Chem.*, **1999**, 7, 39-59.

63 Koepf, E.K.; Petrassi, H.M.; Sudol, M.; Kelly, J.W. “WW: An isolated three-stranded antiparallel β-sheet domain that unfolds and refolds reversibly; Evidence for a structured hydrophobic cluster in urea and GdnHCl and a disordered thermal unfolded state” *Protein Sci.*, **1999**, 8, 841-853.

62 Lashuel, H.A.; Lai, Z.; Kelly, J.W. "Characterization of the Transthyretin Acid Denaturation Pathways by Analytical Ultracentrifugation: Implications for Wild Type, V30M and L55P Amyloid fibril Formation, *Biochemistry*, **1998**, 37, 17851-17864.

61 Angeletti, R.H.; Bibbs, L.; Bonewald, L.F.; Fields, G.B.; Kelly, J.W.; McMurray, J.S.; Moore, W.T.; Weintraub, S.T. Proc. *Eur. Pept. Symp.*, 24th (**1998**) 207-208.

60 Kelly, J.W. “Overview of the Biosynthesis of Amino Acids, Peptides, Porphyrins and Alkaloids with a Focus on the Biosynthesis of Aromatic Amino acids” Comprehensive Natural Products Chemistry, Vol 4, Amino Acids, Peptides, Porphyrins and Alkaloids. Kelly, J.W. Ed., Elsevier Science Ltd, Oxford, **1998**, 1-11.

59 Peterson, S.A.; Klabunde, T.; Lashuel, H.A.; Purkey, H.; Sacchettini, J.C.; Kelly, J.W. "Inhibiting Transthyretin Conformational Changes That Lead To Amyloid Fibril Formation" *Proc. Natl. Acad. Sci USA*, **1998**, 95, 12956-12960.

58 Nettleton, E.J.; Sunde, M.; Lai, Z.; Kelly, J.W.; Dobson, C.M.; Robinson, C.V. “Protein Subunit Interactions and Structural Integrity of Amyloidogenic Transthyretins-Evidence from Electrospray Mass Spectrometry” *J. Mol. Biol.* **1998**, 281, 553-564.

57 Xie, Y.; Laushel, H.A.; Miroy, G.J.; Dikler, S.; Kelly, J.W. “Recombinant Human Retinol-Binding Protein Refolding, Native Disulfide Formation and Characterization” *Protein Expression and Purification* **1998**, 14, 31-37.

56 LaBrenz, S.R.; Bekele, H.; Kelly, J.W. "Enhancement of the Water Solubility of Aromatic Molecules Via the Heck Reaction. A Comparison of ethano-Ammonium, -Carboxylate, and -Phosphonate Functional Groups" *Tetrahedron* **1998**, 54, 8671-8678.

55 Patricelli, M.P.; Lashuel, H.A.; Giang, D.K.; Kelly, J.W.; Cravatt, B.F. “Comparative Characterization of a Wild Type and Transmembrane Domain-Deleted Fatty Acid Amide Hydrolase: Identification of the Transmembrane Domain as a Site for Oligomerization” *Biochemistry* **1998**, 37, 15177-15187.

54 Baures, P.W.; Peterson, S.A.; Kelly, J.W. “Discovering Transthyretin Amyloid Fibril Inhibitors by Limited Screening” Bioorg. Med. Chem., **1998**, 6, 1389-1401.

53 Kelly, J.W. “The Alternative Conformations of Amyloidogenic Proteins and Their Multi-Step Assembly Pathways” *Curr. Opin. Struct. Biol.*, **1998**, 8, 101-106.

52 Kelly, J.W. “The Environmental Dependency of Protein Folding Best Explains Prion and Amyloid Disease” *Proc. Natl. Acad. Sci. USA*, **1998**, 95, 930-932.

51 Dikler, S.; Kelly, J.W.; Russell, D.H. "Improving mass spectrometry sequencing of arginine-containing peptides by derivitization with acetylacetone" *J. Mass. Spectrom.* **1997**, 32, 1337-1349.

50 Kelly, J.W. "Amyloid Fibril Formation and protein misassembly: a structural quest for insights into amyloid and prion diseases", *Structure* **1997**, 5, 595-600.

49 Lai, Z.; McCulloch, J.;Laushel. H.A. Kelly, J.W. "Guanidine hydrocholoride–Induced Denaturation and Refolding of Transthyretin Exhibits A Marked Hysteresis: Equilibria With High Kinetic Barriers, *Biochemistry*, **1997**, 36, 10230-10239.

48 Kelly, J.W.; Colon, W.; Lai, V.; Lashuel, H.; McVulloch, J.; McCutchen, S.; Miroy, G.; Peterson, S.; "Transthyretin Quaternary and Tertiary Structural Changes Facilitate Misassembly into Amyloid, *Advances in Protein Chemistry*, **1997**, 50, 161-181.

47 Bekele, H.; Nesloney, C.L.; McWilliams, K.W.; Zacharias, N.M.; Chitnumsub, P.; Kelly, J.W. Improved Synthesis of the Boc and Fmoc Derivatives of 4-(2'–Aminoethyl)-6-Dibenzofuranpropionic acid: An Unnatural Amino Acid That Nucleates β-Sheet Folding. *J. Org. Chem.* **1997**, 62, 2259-2262.

46 Chen, H.I.; Einbond, A.; Kwak, S-J; Linn, H.; Koepf, E.;Peterson, S.; Kelly, J.W.; Sudol, M. "Charaterization of the WW domain of human Yes-associated protein and its polyproline-containing ligands, *J. Biol. Chem.* **1997**, 272, 17070-17077.

45 Zhang, W.; Diaz, H.; Kelly, J.W.; Taylor, J.W. "Examining the Structural and Functional Roles of the β-Turn Region in Neuropeptide Y *Lett. Pep. Sci.* **1996**, 3, 133-142.

44 Miroy, G.J.; Lai Z.; Lashuel, H.A.; Peterson, S.A. Strang, C.J.; Kelly J.W. "Inhibiting Transthyretin Amyloid Fibril Formation Via Protein Stabilization" *Proc. Natl. Acad. Sci. USA*, **1996**, 93, 15051-15056.

43 McWilliams, K.; Kelly, J.W. Synthesis and Conformational Preferences of a Potential β-Sheet Nucleator Based on the 9,9-Dimethylxanthene Skeleton *J. Org. Chem.* **1996**, 61, 7408-7414.

42 Colon, W.; Lai, Z.; McCutchen, S.L.; Miroy, G.J.; Strang, C.J.; Kelly, J.W. 1996 " FAP Mutations Destabilize Transthyretin Facilitating Conformational Changes Required For Amyloid Formation" In: “The Nature and Origin of Amyloid Fibrils” (CIBA Foundation Symposium 1996) Wiley, Chichester, **1996**, 228-242.

41 Nesloney, C. & Kelly, J.W. "Progress Towards Understanding β-Sheet Structure" *Bioorganic and Medicinal Chemistry*, **1996**, 4, 739-766.

40 Nesloney, C.N.; Kelly, J.W. "A 2,3'-Substituted Biphenyl Based Amino Acid Facilitates The Formation Of A Monomeric β-Hairpin-Like Structure In Aqueous Solution at Elevated Temperature" *J. Am. Chem. Soc.*, **1996**, 118, 5836-5845.

39 Nesloney, C.L.; Kelly, J.W. "Synthesis and Hydrogen Bonding Capabilities of Bipheyl-Based Amino Acids Designed To Nucleate β-Sheet Structure" *J. Org. Chem*. **1996**, 61, 3127-3137.

38 Lai, Z.; Colon, W. ; Kelly, J.W. "The Acid-Mediated Denaturation Pathway of Transthyretin Yields A Conformational Intermediate Which Can Self-Assemble Into Amyloid" *Biochemistry*, **1996**, 35, 6470-6482.

37 Kelly, J.W. "Alternative Conformations of Amyloidogenic Proteins Govern Their Behavior" *Curr. Op. Struct. Biol.*, **1996**, 6, 11-17.

36 Choo, D; Schneider, J.P.; Graciani, N.R.; Kelly, J.W. "Nucleated Antiparallel β-Sheet That Folds and Undergoes Self-Assembly: A Template Promoted Folding Strategy Toward Controlled Molecular Architectures" *Macromolecules*, **1996**, 29, 355-366.

35 McCutchen, S.L.; Lai, Z.; Miroy, G.J.; Kelly, J.W.; Colon, W. "Comparison of Lethal and Non-Lethal Transthyretin Variants and Their Relationship To Amyloid Disease" *Biochemistry*, **1995**, 34, 13527-13536.

34 Schneider, J.; Kelly, J.W."Templates That Induce α-Helical, β-Sheet and Loop Conformations" *Chemical Reviews*. **1995**, 95, 2169-2187.

33 Schneider, J.; Kelly, J.W." Synthesis and Efficacy of Square Planar Copper Complexes Designed To Nucleate β-Sheet Secondary Structure" *J. Am. Chem. Soc.*, **1995**, 117, 2533-2546.

32 Graciani, N.;Swanson, D.T.; Kelly, J.W. "Phosphomonoesters and Phosphodiesters Derived From the Photohydrolysis of 2-methoxy-5-Nitropheny Substituted Phosphotriesters"*Tetrahedron.* **1995**, 51, 1077-1086.

31 LaBrenz, S.R.; Kelly, J.W. "A Peptidomimetic Host That Binds A Peptide Guest Affording a β-Sheet Structure that Subsequently Self-Assembles; A simple Receptor Mimic" *J. Am. Chem. Soc.*, **1995**, *117*, 1655-1656.

30 Graciani, N.R.; Tsang, K.Y.; McCutchen, S.L.; Kelly, J.W. "Amino Acids That Specify Structure Through Hydrophobic Clustering And Histidine-Aromatic Interactions Lead To Biologically Active Peptidomimetics" *Bioorganic and Med. Chem.* **1994**, *2*, 999-1006.

29 Kelly, J.W.; Lansbury. P.T.. "A Chemical Approach to Elucidate the Mechanism of Transthyretin and β-Protein Amyloid Fibril Formation" *Amyloid*, **1994**, *1*, 186-205.

28 Tsang, K-Y.; Diaz, H.; Graciani, N.; Kelly, J.W. "Hydrophobic Cluster Formation is Necessary for Dibenzofuran-Based Amino Acids to Function as β-Sheet Nucleators" *J. Am. Chem. Soc.* **1994**, *116*, 3988-4005.

27 McCutchen, S.L.; Kelly, J.W. "Intermolecular Disulfide Linkages Are Not Required For Transthyretin Amyloid Fibril Formation *In Vitro*" *Biochem. Biophys. Res. Comm.* **1993**, *197*, 415-421.

26 McCutchen, S.; Colon, W.; Kelly, J.W. "Transthyretin Mutation Leu-55-Pro Significantly Alters Tetramer Stability and Increases Amyloidogenicity" *Biochemistry* **1993**, *32*, 12119-12127.

25 Diaz, H.; Tsang, K.Y.; Choo, D. Kelly, J.W. "The Design of Water Soluble β-Sheet Structure Based on a Nucleation Strategy" *Tetrahedron* **1993**, *49*, 3533 - 3545.

24 Diaz, H..; Tsang, K.Y.; Choo, D.; Espina, J.R.; Kelly, J.W. "Design, Synthesis And Partial Characterization Of Water–Soluble β-Sheets Stabilized By a Dibenzofuran–Based Amino Acid" *J. Am. Chem. Soc.* **1993**, 115, 3790-3791.

23 Diaz, H.; Espina, J. R.; Kelly, J.W. "A Dibenzofuran Based Amino Acid Designed To Nucleate Antiparallel β-Sheet Structure: Evidence For Intramolecular Hydrogen Bond Formation*" J. Am. Chem. Soc.* **1992**, 114, 8316-8318.

22 Colon, W.; Kelly, J.W. "Partial Denaturation of Transthyretin is Sufficient for Amyloid Fibril Formation In Vitro." *Biochemistry*, **1992**, 31, 8654-8660.

21 Banzon, J.A.; Kelly, J.W. "β-Sheet Rearrangements: Serpins and Beyond" *Prot. Engineering* **1992**, 5, 113-115.

20 Diaz, H.; Kelly, J.W. "The Synthesis of Dibenzofuran Based Diacids and Amino Acids Designed To Nucleate Parallel and Antiparallel β-sheet Formation" *Tetrahedron Lett.* **1991**, 32, 5725-5728.

19 Colon, W.; Kelly, J.W. "Transthyretin Acid Induced Denaturation is Required For Amyloid Fibril Formation in vitro." in "*Applications of Enzyme Biotechnology*" Kelly, J.W. & Balwin, T.O. Eds; Plenum: New York, **1991**, pp 99-108.

18 Schneider, J.P.; Topgi, R.S., Kelly, J.W. "A Convenient Synthesis of 6,6'-Diamino-2,2'-Bipyridine" *Synthetic Comm.* **1992**, 22, 1033-1037.

17 Kelly, J.W.; Robinson, P.L., Murray, W.T.; Eskew, N.A.; Pautard Cooper, A.; Mathieu- P.; Evans, S.A., Jr. "New Synthetic Methodologies Employing Substituted 1,3,2,5-Dioxaphospholanes" *ACS Symposium Series* **1992**, 486, 186-201.

16 Kaiser, E. T.; Mihara, H.; Laforet, G. A.; Kelly J.W.; Walters, L.; Findeis, M.A. Sasaki, T. "Peptide and Protein Synthesis by Segment Synthesis-Condensation" *Science*. **1989**, 243, 187-192.

15 Kelly, J. W. "New Tools for Probing Protein Structure" *Bio/Technology* **1988**, 6, 125-129.

14 Kelly, J.W.; Walters, L.; Kaiser, E.T. "The Oxime Based Segment Synthesis of Cro" in "The Analysis of Peptides and Proteins by Mass Spectrometry", C.J. McNeal, editor. Wiley, 1988, 15-19.

13 Scheim, U.; Ruhlmann, K.; Evans, S.A., Jr.; Kelly, J.W.; Bassindale, A.R. "Synthesis of Siloxanes: Oxygen-17 NMR Spectroscopy" *J. Organometallic Chem.* **1988**, 340, 19-21.

12 Kelly, J. W.; Evans, S.A., Jr. "Oxygen-17 NMR Spectral Studies of Selected Aromatic Sulfones" *Mag. Res. Chem.* **1987**, 25, 305-308.

11 Kelly, J. W.; Robinson, P.L.; Evans, S.A., Jr. "Dioxyphosphoranes, an Emerging Class of Versatile Reagents for Organic Synthesis" *Phosphorus and Sulfur*. **1987**, 30, 147-150.

10 Murray, W. T.; Kelly, J.W.; Evans, S.A., Jr. "Synthesis of Substituted 1,4- Oxathianes Mechanistic Details of Diethoxytriphenylphosphorane- and Triphenylphosphine / Tetrachloromethane-Promoted Cyclodehydrations and 13C NMR Spectroscopy" *J. Org. Chem.* **1987**, 52, 525-529.

9 Robinson, P. L.; J. W. Kelly; Evans, S.A. "The Chemistry of Mercaptoalcohols in the Presence of Diethoxytriphenylphosphorane Temperature Dependence of Cyclodehydrations and S-Ethylations" *Phosphorus and Sulfur* **1987**, 31, 59-70.

8 Kelly, J. W.; Eskew, N.L.; Evans, S.A., Jr. "Cyclohydration of N- and C-Substituted β-amino Alcohols to the Corresponding Aziridines with Diethoxytriphenylphosphorane" *J. Org. Chem.* **1986**, 51, 95-97.

7 Kelly, J. W.; Evans, S.A., Jr. "Bis (neopentyloxy) triphenylphosphorane: A Versatile, Nonalkylating Cyclodehydration Reagent" *J. Org. Chem.* **1986**, 51, 5490-5492.

6 Kelly, J. W.; Evans, S.A., Jr. "Regioselective Phosphoranylation and Cyclodehydration of Triols with Diethoxytriphenylphosphorane" *J. Am. Chem. Soc.* **1986**, 108, 7681-7685.

5 Kelly, J. W.; Robinson, P.L.; Evans, S.A., Jr. "2,2,2-Triphenyl-4,5-(2',2"-biphenylene)-1,3,2-dioxaphosphane: An Effective Reagent for Converting Diols, Amino Alcohols and Especially Mercapto Alcohol to the Corresponding Heterocycles" *J. Org. Chem.* **1986**, 51, 4473-4475.

4 Robinson, P. L.; Kelly, J.W.; Evans, S. A., Jr. "The Synthetic Utility of Dioxyphosphoranes in Organic Synthesis" *Phosphorus and Sulfur* **1986**, 26, 15-24.

3 "1. The Mechanism, Stereochemistry, and Scope of Dioxyphosphorane Promoted Cyclodehydrations. 2. Oxygen-17 NMR Studies Directed Towards Elucidating The Structure And Dynamics Of Oxygenated Organosulfur and Organosilicon Compounds" Kelly, J.W. Ph.D. Dissertation, University of North Carolina at Chapel Hill, N.C., 1986.

2 Robinson, P. L.; Barry, C.N.; Kelly, J.W., Evans, S.A., Jr. "Diethoxytriphenylphosphorane: A Mild Regioselective Cyclodehydrating Reagent for Conversion of Diols to Cyclic Ethers Stereochemistry, Synthetic Utility and Scope"
J. Am. Chem. Soc. **1985**, 107, 5210-5219.

1 Kelly, J. W., P. L. Robinson and S. A. Evans. "Diethoxydiphenylpolystyrylphosphorane: A New Polymeric Reagent for the Efficient Cyclodehydration of Simple Diols" *J. Org. Chem.* **1985**, 50, 5007-5009.

Patents

Title	Inventor(s)	US Patent #	Patent Date
Regulators Of The Endoplasmic Reticulum Proteostasis Network	C. Cooley J.W. Kelly R. Paxman L. Plate R.L. Wiseman	PCT Pending	Date of Application 7/6/2017
Sulfur(Vi) Fluoride Compounds and Methods For The Preparation Thereof	J. Dong K.B. Sharpless J.W. Kelly W. Chen A. Baranczak	WO 15/188120 PCT published	Date of Application 6/5/2015
Transthyretin Antibodies and Uses Thereof	X. Jiang J.W. Kelly J. Chapman	PCT Pending	Publication Date 2/11/2016
Small Molecules That Covalently Modify Transthyretin	J.W. Kelly S. Choi	8703815	4/22/2014
Reliable Stabilization Of N-Linked Polypeptide Native States With Enhanced Aromatic Sequons Located In Polypeptide Tight Turns	J.W. Kelly J.L. Price E.K. Culyba E.T. Powers	8906681	12/9/2014
Inhibitors Of Transthyretin Amyloid Fibril Formation	J.W. Kelly S.M. Johnson H.M. Petrassi	7312361	12/25/2007 Expired
Transthyretin Stabilization	J.W. Kelly H.M. Petrassi	WO 05/112913 PCT	Published 12/2005 Archived 6/2011
Methods For Identifying Cellular Modulators Of Disaggregation Activity Or Aggregation Activity In An Animal	J.W. Kelly J.G. Bieschke A. Dillin E. Cohen	WO 08/022035 PCT	Published 2/2008 Archived 5/2015
Genistein Inhibition Of Transthyretin Amyloidosis	J.W. Kelly N.S. Green	WO 07/022138 PCT	Published 2/2007 Archived 7/2010
Compounds, Compositions And Methods For Stabilizing Transthyretin And Inhibiting Transthyretin Misfolding	J.W. Kelly	PCT published	12/2005
Compositions and Methods For Stabilizing Transthyretin and Inhibiting Transthyretin Misfolding	J.W. Kelly Y. Sekijima	7560488 7214696	7/14/2009 5/8/2007
Compositions and Methods For Stabilizing Transthyretin and Inhibiting Transthyretin Misfolding	J.W. Kelly E.T. Powers H. Razavi	8653119 7214695	2/18/2014 5/8/2007
Methods For Treating A Condition Characterized By Dysfunction In Protein Homeostasis	L. Segatori T. Mu J.W. Kelly	WO 09/100037 PCT	Published 8/2009 Archived 5/2015
Chemical Chaperones And Their Effect Upon The Cellular Activity Of Beta-Glucosidase	J.W. Kelly A.R. Sawkar E. Beutler C-H. Wong W.E. Balch	WO 04/037373 PCT	Published 05/2004 Archived 9/2008
See next page for table continuation Anti-amyloidogenic agents	 G. Miroy	 WO9827972 (A2)	 7/2/1998

	J.W. Kelly Z. Lai H. Lashuel S.A. Peterson	WO9827972 (A3) WO9827972 (A8)	
Beta -Sheet Nucleating Peptidomimetics	J.W. Kelly	6034211	March 7, 2000