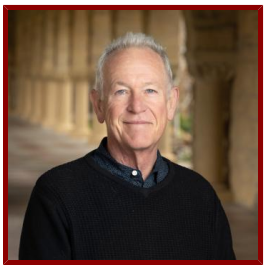
Stanford



Ron Kopito

Professor of Biology

Curriculum Vitae available Online

CONTACT INFORMATION

- **Administrative Contact**

  Kelly Cosmas - Adminstrative Assistant

  Email kcosmas@stanford.edu

  Tel 650-725-8777

Bio

ACADEMIC APPOINTMENTS

- Professor, Biology
- Member, Bio-X
- Member, Maternal & Child Health Research Institute (MCHRI)
- Member, Wu Tsai Neurosciences Institute

HONORS AND AWARDS

- Lifetime Fellow, American Society fo Cell Biology (2018)
- Basil O'Connor Award, March of Dimes (1989)
- Scholar in Biomedical Science, Lucille P. Markey Foundation (1985)
- Presidential Young Investigator, National Science Foundation (1989)
- Established Investigator, American Heart Association (1993)

PROFESSIONAL EDUCATION

- A.B., Bowdoin College , Biochemistry (1976)
- Ph.D., MIT , Biochemistry (1982)

LINKS

- Kopito Lab: https://www.kopitolab.com/

Research & Scholarship

CURRENT RESEARCH AND SCHOLARLY INTERESTS

The ER is the "port of entry" for proteins destined for the cell surface and beyond. The vast majority of proteins entering the secretory pathway are synthesized on ribosomes docked at ER translocons and are co-transationally translocated into the ER lumen. Proteins synthesized at the ER are subject to covalent modifications that include N- and O-glycosylation, disulfide bond formation, and in some cases, proline and lysine hydroxylation.

Membrane proteins must be threaded co-translocationally into the lipid bilayer to become membrane-integrated, often with complex topologies and typically form hetero- or homo- oligomers. This highly complex "protein biogenesis" process is assisted by a diverse network of folding catalysts and protein-modifying enzymes and is scrutinized by molecular chaperones and other "quality control" factors which ensure that only correctly folded and assembled proteins exit the ER and proceed to distal compartments of the secretory pathway.

The Kopito laboratory seeks a molecular understanding of how cells maintain the fidelity of their proteomes. Unlike DNA, which can be repaired if damaged or incorrectly made, proteins cannot be mended. Instead, damaged or incorrectly synthesized proteins must be rapidly and efficiently destroyed lest they form toxic aggregates.

Our goal is to elucidate the functional networks that coordinate protein synthesis and quality control in the early secretory pathway. Currently the lab is focused on two specific systems: ERAD and ribosome UFMylation.

Teaching

COURSES

2026-27

- Biology PhD Lab Rotation: BIO 299 (Aut, Win, Spr)
- Cell Biology: BIO 86 (Spr)
- Cystic fibrosis: from medical conundrum to precision medicine success story: BIO 25N (Win)

2025-26

- Cell Biology: BIO 86 (Spr)

2024-25

- Advanced Cell Biology: BIO 214, BIOC 224, MCP 221 (Win)
- Cell Biology: BIO 86 (Spr)
- Cystic fibrosis: from medical conundrum to precision medicine success story: BIO 25Q (Win)

2023-24

- Advanced Cell Biology: BIO 214, BIOC 224, MCP 221 (Win)
- Cell Biology: BIO 86 (Spr)
- Cystic fibrosis: from medical conundrum to precision medicine success story: BIO 25Q (Win)

STANFORD ADVISEES

Doctoral Dissertation Reader (AC)

Pre Lavania, Angel Madero Rincon, Caroline Scheuing, Ananya Vinayak

Postdoctoral Faculty Sponsor

Constantine Bartolutti, Magda Wachalska, Jingyi Zhang

Doctoral Dissertation Advisor (AC)

Carlo Armijo

GRADUATE AND FELLOWSHIP PROGRAM AFFILIATIONS

- Biochemistry (PhD Program)
- Biology (School of Humanities and Sciences) (PhD Program)

- Biophysics (PhD Program)
- Neurosciences (PhD Program)

Publications

PUBLICATIONS

- **Evaluating evidence for UFMylation client diversity.** *Nature reviews. Molecular cell biology*
  Scavone, F., Kopito, R. R.
  2026
- **UFMylation orchestrates spatiotemporal coordination of RQC at the ER.** *Science advances*
  Penchev, I., Gumbin, S., Scavone, F., Berninghausen, O., Becker, T., Kopito, R., Beckmann, R.
  2025; 11 (18): eadv0435
- **UFM1 E3 ligase promotes recycling of 60S ribosomal subunits from the ER.** *Nature*
  DaRosa, P. A., Penchev, I., Gumbin, S. C., Scavone, F., Wąchalska, M., Paulo, J. A., Ordureau, A., Peter, J. J., Kulathu, Y., Harper, J. W., Becker, T., Beckmann, R., Kopito, et al
  2024
- **Small molecule correctors divert CFTR-F508del from ERAD by stabilizing sequential folding states.** *Molecular biology of the cell*
  Riepe, C., Wąchalska, M., Deol, K. K., Amaya, A. K., Porteus, M. H., Olzmann, J. A., Kopito, R. R.
  2023: mbcE23080336
- **RPL26/uL24 UFMylation is essential for ribosome-associated quality control at the endoplasmic reticulum.** *Proceedings of the National Academy of Sciences of the United States of America*
  Scavone, F., Gumbin, S. C., Da Rosa, P. A., Kopito, R. R.
  2023; 120 (16): e2220340120
- **Ribosomal protein RPL26 is the principal target of UFMylation.** *Proceedings of the National Academy of Sciences of the United States of America*
  Walczak, C. P., Leto, D. E., Zhang, L., Riepe, C., Muller, R. Y., DaRosa, P. A., Ingolia, N. T., Elias, J. E., Kopito, R. R.
  2019
- **Acute unfolding of a single protein immediately stimulates recruitment of ubiquitin protein ligase E3C (UBE3C) to 26S proteasomes.** *The Journal of biological chemistry*
  Gottlieb, C. D., Thompson, A. C., Ordureau, A. n., Harper, J. W., Kopito, R. R.
  2019
- **Genome-wide CRISPR Analysis Identifies Substrate-Specific Conjugation Modules in ER-Associated Degradation.** *Molecular cell*
  Leto, D. E., Morgens, D. W., Zhang, L., Walczak, C. P., Elias, J. E., Bassik, M. C., Kopito, R. R.
  2018
- **Redundant and Antagonistic Roles of XTP3B and OS9 in Decoding Glycan and Non-glycan Degrons in ER-Associated Degradation** *MOLECULAR CELL*
  van der Goot, A. T., Pearce, M. M. P., Leto, D. E., Shaler, T. A., Kopito, R. R.
  2018; 70 (3): 516-+
- **Spatial regulation of UBXD8 and p97/VCP controls ATGL-mediated lipid droplet turnover** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
  Olzmann, J. A., Richter, C. M., Kopito, R. R.
  2013; 110 (4): 1345-1350
- **Defining human ERAD networks through an integrative mapping strategy** *NATURE CELL BIOLOGY*
  Christianson, J. C., Olzmann, J. A., Shaler, T. A., Sowa, M. E., Bennett, E. J., Richter, C. M., Tyler, R. E., Greenblatt, E. J., Harper, J. W., Kopito, R. R.
  2012; 14 (1): 93-U176
- **Prion-like transmission of protein aggregates in neurodegenerative diseases** *NATURE REVIEWS MOLECULAR CELL BIOLOGY*
  Brundin, P., Melki, R., Kopito, R.

2010; 11 (4): 301-307

- **Cytoplasmic penetration and persistent infection of mammalian cells by polyglutamine aggregates** *NATURE CELL BIOLOGY*
  Ren, P., Lauckner, J. E., Kachirskaia, I., Heuser, J. E., Melki, R., Kopito, R. R.
  2009; 11 (2): 219-U232
- **Hypothalamic neurodegeneration and adult-onset obesity in mice lacking the Ubb polyubiquitin gene** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
  Ryu, K., Garza, J. C., Lu, X., Barsh, G. S., Kopito, R. R.
  2008; 105 (10): 4016-4021
- **OS-9 and GRP94 deliver mutant alpha 1-antitrypsin to the Hrd1-SEL1L ubiquitin ligase complex for ERAD** *NATURE CELL BIOLOGY*
  Christianson, J. C., Shaler, T. A., Tyler, R. E., Kopito, R. R.
  2008; 10 (3): 272-U13
- **Global changes to the ubiquitin system in Huntington's disease** *NATURE*
  Bennett, E. J., Shaler, T. A., Woodman, B., Ryu, K., Zaitseva, T. S., Becker, C. H., Bates, G. P., Schulman, H., Kopito, R. R.
  2007; 448 (7154): 704-U11
- **Central pore residues mediate the p97/VCP activity required for ERAD** *MOLECULAR CELL*
  DeLaBarre, B., Christianson, J. C., Kopito, R. R., Brunger, A. T.
  2006; 22 (4): 451-462
- **HDAC6 and microtubules are required for autophagic degradation of aggregated Huntingtin** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Iwata, A., Riley, B. E., Johnston, J. A., Kopito, R. R.
  2005; 280 (48): 40282-40292
- **Global impairment of the ubiquitin-proteasome system by nuclear or cytoplasmic protein aggregates precedes inclusion body formation** *MOLECULAR CELL*
  Bennett, E. J., Bence, N. F., Jayakumar, R., Kopito, R. R.
  2005; 17 (3): 351-365
- **Impairment of the ubiquitin-proteasome system by protein aggregation** *SCIENCE*
  Bence, N. F., Sampat, R. M., Kopito, R. R.
  2001; 292 (5521): 1552-1555
- **Temporal control of acute protein aggregate turnover by UBE3C and NRF1-dependent proteasomal pathways.** *Proceedings of the National Academy of Sciences of the United States of America*
  Hickey, K. L., Panov, A., Whelan, E. M., Schäfer, T., Mizrak, A., Kopito, R. R., Baumeister, W., Fernández-Busnadiego, R., Harper, J. W.
  2024; 121 (50): e2417390121
- **The herpesvirus UL49.5 protein hijacks a cellular C-degron pathway to drive TAP transporter degradation.** *Proceedings of the National Academy of Sciences of the United States of America*
  Wąchalska, M., Riepe, C., Ślusarz, M. J., Graul, M., Borowski, L. S., Qiao, W., Foltyńska, M., Carette, J. E., Bieńkowska-Szewczyk, K., Szczesny, R. J., Kopito, R. R., Lipińska, A. D.
  2024; 121 (11): e2309841121
- **Small molecule correctors divert CFTR-F508del from ERAD by stabilizing sequential folding states.** *bioRxiv : the preprint server for biology*
  Riepe, C., Wachalska, M., Deol, K. K., Amaya, A. K., Porteus, M. H., Olzmann, J. A., Kopito, R. R.
  2023
- **Parallel CRISPR-Cas9 screens identify mechanisms of PLIN2 and lipid droplet regulation.** *Developmental cell*
  Roberts, M. A., Deol, K. K., Mathiowetz, A. J., Lange, M., Leto, D. E., Stevenson, J., Hashemi, S. H., Morgens, D. W., Easter, E., Heydari, K., Nalls, M. A., Bassik, M. C., Kampmann, et al
  2023
- **A non-canonical scaffold-type E3 ligase complex mediates protein UFMylation.** *The EMBO journal*
  Peter, J. J., Magnussen, H. M., DaRosa, P. A., Millrine, D., Matthews, S. P., Lamoliatte, F., Sundaramoorthy, R., Kopito, R. R., Kulathu, Y.
  2022: e111015
- **PEX19 Coordinates Neutral Lipid Storage in Cells in a Peroxisome-Independent Fashion.** *Frontiers in cell and developmental biology*

Lyschik, S., Lauer, A. A., Roth, T., Janitschke, D., Hollander, M., Will, T., Hartmann, T., Kopito, R. R., Helms, V., Grimm, M. O., Schrul, B.
2022; 10: 859052

- **Phagocytic glia are obligatory intermediates in transmission of mutant huntingtin aggregates across neuronal synapses.** *eLife*
  Donnelly, K. M., DeLorenzo, O. R., Zaya, A. D., Pisano, G. E., Thu, W. M., Luo, L. n., Kopito, R. R., Panning Pearce, M. M.
  2020; 9
- **Methods for genetic analysis of mammalian ER-associated degradation** *UBIQUITIN-DEPENDENT PROTEIN DEGRADATION*
  Leto, D. E., Kopito, R. R.
  edited by Hochstrasser, M.
  2019; 619: 97–120
- **Methods for genetic analysis of mammalian ER-associated degradation.** *Methods in enzymology*
  Leto, D. E., Kopito, R. R.
  2019; 619: 97–120
- **Proteomic analysis of monolayer-integrated proteins on lipid droplets identifies amphipathic interfacial alpha-helical membrane anchors.** *Proceedings of the National Academy of Sciences of the United States of America*
  Pataki, C. I., Rodrigues, J., Zhang, L., Qian, J., Efron, B., Hastie, T., Elias, J. E., Levitt, M., Kopito, R. R.
  2018
- **Characterization of protein complexes of the endoplasmic reticulum-associated degradation E3 ubiquitin ligase Hrd1.** *journal of biological chemistry*
  Hwang, J., Walczak, C. P., Shaler, T. A., Olzmann, J. A., Zhang, L., Elias, J. E., Kopito, R. R.
  2017; 292 (22): 9104-9116
- **Prion-Like Characteristics of Polyglutamine-Containing Proteins.** *Cold Spring Harbor perspectives in medicine*
  Pearce, M. M., Kopito, R. R.
  2017
- **Ubiquitin Accumulation on Disease Associated Protein Aggregates Is Correlated with Nuclear Ubiquitin Depletion, Histone De-Ubiquitination and Impaired DNA Damage Response** *PLOS ONE*
  Ben Yehuda, A., Risheq, M., Novoplansky, O., Bersuker, K., Kopito, R. R., Goldberg, M., Brandeis, M.
  2017; 12 (1)
- **Protein misfolding in neurodegenerative diseases: implications and strategies.** *Translational neurodegeneration*
  Sweeney, P., Park, H., Baumann, M., Dunlop, J., Frydman, J., Kopito, R., McCampbell, A., LeBlanc, G., Venkateswaran, A., Nurmi, A., Hodgson, R.
  2017; 6: 6-?
- **Ron R. Kopito: Unfolding the Secrets of Protein Aggregation.** *Trends in cell biology*
  Kopito, R. R.
  2016; 26 (8): 559-560
- **Peroxin-dependent targeting of a lipid-droplet-destined membrane protein to ER subdomains** *NATURE CELL BIOLOGY*
  Schrul, B., Kopito, R. R.
  2016; 18 (7): 740-?
- **Protein misfolding specifies recruitment to cytoplasmic inclusion bodies** *JOURNAL OF CELL BIOLOGY*
  Bersuker, K., Brandeis, M., Kopito, R. R.
  2016; 213 (2): 229-241
- **Prion-like transmission of neuronal huntingtin aggregates to phagocytic glia in the Drosophila brain.** *Nature communications*
  Pearce, M. M., Spartz, E. J., Hong, W., Luo, L., Kopito, R. R.
  2015; 6: 6768-?
- **Prion-like transmission of neuronal huntingtin aggregates to phagocytic glia in the Drosophila brain.** *Nature communications*
  Pearce, M. M., Spartz, E. J., Hong, W., Luo, L., Kopito, R. R.
  2015; 6: 6768-?
- **Heat shock response activation exacerbates inclusion body formation in a cellular model of huntington disease.** *journal of biological chemistry*

Bersuker, K., Hipp, M. S., Calamini, B., Morimoto, R. I., Kopito, R. R.
2013; 288 (33): 23633-23638

- **Simultaneous Measurement of Amyloid Fibril Formation by Dynamic Light Scattering and Fluorescence Reveals Complex Aggregation Kinetics** *PLOS ONE*
  Streets, A. M., Sourigues, Y., Kopito, R. R., Melki, R., Quake, S. R.
  2013; 8 (1)
- **Simultaneous measurement of amyloid fibril formation by dynamic light scattering and fluorescence reveals complex aggregation kinetics.** *PloS one*
  Streets, A. M., Sourigues, Y., Kopito, R. R., Melki, R., Quake, S. R.
  2013; 8 (1)
- **The Mammalian endoplasmic reticulum-associated degradation system.** *Cold Spring Harbor perspectives in biology*
  Olzmann, J. A., Kopito, R. R., Christianson, J. C.
  2013; 5 (9)
- **Unassembled CD147 is an endogenous endoplasmic reticulum-associated degradation substrate** *MOLECULAR BIOLOGY OF THE CELL*
  Tyler, R. E., Pearce, M. M., Shaler, T. A., Olzmann, J. A., Greenblatt, E. J., Kopito, R. R.
  2012; 23 (24): 4668-4678
- **ALIX Is a Lys63-Specific Polyubiquitin Binding Protein that Functions in Retrovirus Budding** *DEVELOPMENTAL CELL*
  Dowlatshahi, D. P., Sandrin, V., Vivona, S., Shaler, T. A., Kaiser, S. E., Melandri, F., Sundquist, W. I., Kopito, R. R.
  2012; 23 (6): 1247-1254
- **Making the cut: intramembrane cleavage by a rhomboid protease promotes ERAD** *NATURE STRUCTURAL & MOLECULAR BIOLOGY*
  Greenblatt, E. J., Olzmann, J. A., Kopito, R. R.
  2012; 19 (10): 979-981
- **Fibrillar Structure and Charge Determine the Interaction of Polyglutamine Protein Aggregates with the Cell Surface** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Trevino, R. S., Lauckner, J. E., Sourigues, Y., Pearce, M. M., Bousset, L., Melki, R., Kopito, R. R.
  2012; 287 (35): 29722-29728
- **Indirect inhibition of 26S proteasome activity in a cellular model of Huntington's disease** *JOURNAL OF CELL BIOLOGY*
  Hipp, M. S., Patel, C. N., Bersuker, K., Riley, B. E., Kaiser, S. E., Shaler, T. A., Brandeis, M., Kopito, R. R.
  2012; 196 (5): 573-587
- **Perturbation of the Hematopoietic System during Embryonic Liver Development Due to Disruption of Polyubiquitin Gene Ubc in Mice** *PLOS ONE*
  Ryu, K., Park, H., Rossi, D. J., Weissman, I. L., Kopito, R. R.
  2012; 7 (2)
- **Live-cell imaging of ubiquitin-proteasome system function.** *Methods in molecular biology (Clifton, N.J.)*
  Hipp, M. S., Bersuker, K., Kopito, R. R.
  2012; 832: 463-472
- **Defining human ERAD networks through an integrative mapping strategy.** *Nature cell biology*
  Christianson, J. C., Olzmann, J. A., Shaler, T. A., Sowa, M. E., Bennett, E. J., Richter, C. M., Tyler, R. E., Greenblatt, E. J., Harper, J. W., Kopito, R. R.
  2012; 14 (1): 93-105
- **Derlin-1 is a rhomboid pseudoprotease required for the dislocation of mutant alpha-1 antitrypsin from the endoplasmic reticulum** *NATURE STRUCTURAL & MOLECULAR BIOLOGY*
  Greenblatt, E. J., Olzmann, J. A., Kopito, R. R.
  2011; 18 (10): 1147-U115
- **Lipid Droplet Formation Is Dispensable for Endoplasmic Reticulum-associated Degradation** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Olzmann, J. A., Kopito, R. R.
  2011; 286 (32): 27872-27874

- **Protein standard absolute quantification (PSAQ) method for the measurement of cellular ubiquitin pools** *NATURE METHODS*
  Kaiser, S. E., Riley, B. E., Shaler, T. A., Trevino, R. S., Becker, C. H., Schulman, H., Kopito, R. R.
  2011; 8 (8): 691-U129
- **Altered Testicular Gene Expression Patterns in Mice Lacking the Polyubiquitin Gene Ubb** *MOLECULAR REPRODUCTION AND DEVELOPMENT*
  Sinnar, S. A., Small, C. L., Evanoff, R. M., Reinholdt, L. G., Griswold, M. D., Kopito, R. R., Ryu, K.
  2011; 78 (6): 415-425
- **Autophagy inhibition engages Nrf2-p62 Ub-associated signaling** *AUTOPHAGY*
  Riley, B. E., Kaiser, S. E., Kopito, R. R.
  2011; 7 (3): 338-340
- **Ubiquitin accumulation in autophagy-deficient mice is dependent on the Nrf2-mediated stress response pathway: a potential role for protein aggregation in autophagic substrate selection** *JOURNAL OF CELL BIOLOGY*
  Riley, B. E., Kaiser, S. E., Shaler, T. A., Ng, A. C., Hara, T., Hipp, M. S., Lage, K., Xavier, R. J., Ryu, K. Y., Taguchi, K., Yamamoto, M., Tanaka, K., Mizushima, et al
  2010; 191 (3): 537-552
- **Loss of polyubiquitin gene Ubb leads to metabolic and sleep abnormalities in mice** *NEUROPATHOLOGY AND APPLIED NEUROBIOLOGY*
  Ryu, K., Fujiki, N., Kazantzis, M., Garza, J. C., Bouley, D. M., Stahl, A., Lu, X., Nishino, S., Kopito, R. R.
  2010; 36 (4): 285-299
- **SPFH1 and SPFH2 mediate the ubiquitination and degradation of inositol 1,4,5-trisphosphate receptors in muscarinic receptor-expressing HeLa cells** *BIOCHIMICA ET BIOPHYSICA ACTA-MOLECULAR CELL RESEARCH*
  Wang, Y., Pearce, M. M., Sliter, D. A., Olzmann, J. A., Christianson, J. C., Kopito, R. R., Boeckmann, S., Gagen, C., Leichner, G. S., Roitelman, J., Wojcikiewicz, R. J.
  2009; 1793 (11): 1710-1718
- **The polyubiquitin Ubc gene modulates histone H2A monoubiquitylation in the R6/2 mouse model of Huntington's disease** *JOURNAL OF CELLULAR AND MOLECULAR MEDICINE*
  Bett, J. S., Benn, C. L., Ryu, K., Kopito, R. R., Bates, G. P.
  2009; 13 (8B): 2645-2657
- **Misfolded proteins partition between two distinct quality control compartments** *NATURE*
  Kaganovich, D., Kopito, R., Frydman, J.
  2008; 454 (7208): 1088-U36
- **The mouse polyubiquitin gene Ubb is essential for meiotic progression** *MOLECULAR AND CELLULAR BIOLOGY*
  Ryu, K., Sinnar, S. A., Reinholdt, L. G., Vaccari, S., Hall, S., Garcia, M. A., Zaitseva, T. S., Bouley, D. M., Boekelheide, K., Handel, M. A., Conti, M., Kopito, R. R.
  2008; 28 (3): 1136-1146
- **The mouse polyubiquitin gene UbC is essential for fetal liver development, cell-cycle progression and stress tolerance** *EMBO JOURNAL*
  Ryu, K., Maehr, R., Gilchrist, C. A., Long, M. A., Bouley, D. M., Mueller, B., Ploegh, H. L., Kopito, R. R.
  2007; 26 (11): 2693-2706
- **Impaired post-translational folding of familial ALS-linked Cu, Zn superoxide dismutase mutants** *EMBO JOURNAL*
  Bruns, C. K., Kopito, R. R.
  2007; 26 (3): 855-866
- **Cellular mechanisms of protein quality control.** *Rinsho shinkeigaku = Clinical neurology*
  Bennett, E. J., Shaler, T., Gonzalez-Zulueta, M., Schulman, H. F., Iwata, A., Riley, B. E., Johnston, J. A., Bucci, M., Nukina, N., Ellerby, L., Kopito, R. R.
  2006; 46 (11): 805-?
- **Cellular mechanisms of protein quality control**
  Bennett, E. J., Shaler, T., Gonzalez-Zulueta, M., Schulman, H. F., Iwata, A., Riley, B. E., Johnston, J. A., Bucci, M., Nukina, N., Ellerby, L., Kopito, R. R.
  LANDES BIOSCIENCE.2006: 344–44

- **Ubiquitin-specific protease 2 as a tool for quantification of total ubiquitin levels in biological specimens** *ANALYTICAL BIOCHEMISTRY*
  Ryu, K., Baker, R. T., Kopito, R. R.
  2006; 353 (1): 153-155

- **Intersecting pathways to neurodegeneration in Parkinson's disease: Effects of the pesticide rotenone on DJ-1, alpha-synuclein, and the ubiquitin-proteasome system** *NEUROBIOLOGY OF DISEASE*
  Betarbet, R., Canet-Aviles, R. A., Sherer, T. B., Mastroberardino, P. G., McLendon, C., Kim, J. H., Lund, S., Na, H. M., Taylor, G., Bence, N. F., Kopito, R., Seo, B. B., Yagi, et al
  2006; 22 (2): 404-420

- **Increased susceptibility of cytoplasmic over nuclear polyglutamine aggregates to autophagic degradation** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
  Iwata, A., Christianson, J. C., Bucci, M., Ellerby, L. M., Nukina, N., Forno, L. S., Kopito, R. R.
  2005; 102 (37): 13135-13140

- **Formation of morphologically similar globular aggregates from diverse aggregation-prone proteins in mammalian cells** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
  Mukai, H., Isagawa, T., Goyama, E., Tanaka, S., Bence, N. F., Tamura, A., Ono, Y., Kopito, R. R.
  2005; 102 (31): 10887-10892

- **Subversion of cellular autophagosomal machinery by RNA viruses** *PLOS BIOLOGY*
  Jackson, W. T., Giddings, T. H., Taylor, M. P., Mulinyawe, S., RABINOVITCH, M., Kopito, R. R., Kirkegaard, K.
  2005; 3 (5): 861-871

- **Effect of ubiquitin expression on neuropathogenesis in a mouse model of familial amyotrophic lateral sclerosis** *NEUROPATHOLOGY AND APPLIED NEUROBIOLOGY*
  Gilchrist, C. A., Gray, D. A., Stieber, A., GONATAS, N. K., Kopito, R. R.
  2005; 31 (1): 20-33

- **Suppression of wild-type rhodopsin maturation by mutants linked to autosomal dominant retinitis pigmentosa** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Rajan, R. S., Kopito, R. R.
  2005; 280 (2): 1284-1291

- **Application and analysis of the GFP(u) family of ubiquitin-proteasome system reporters** *UBIQUITIN AND PROTEIN DEGRADATION, PT B*
  Bence, N. F., Bennett, E. J., Kopito, R. R.
  2005; 399: 481-490

- **The missing linker: An unexpected role for a histone deacetylase** *MOLECULAR CELL*
  Kopito, R. R.
  2003; 12 (6): 1349-1351

- **Immunoglobulin light chains dictate vesicular transport-dependent and -independent routes for IgM degradation by the ubiquitin-proteasome pathway** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Elkabetz, Y., Kerem, A., Tencer, L., Winitz, D., Kopito, R. R., Bar-Nun, S.
  2003; 278 (21): 18922-18929

- **Recognition of a single transmembrane degron by sequential quality control checkpoints** *MOLECULAR BIOLOGY OF THE CELL*
  Fayadat, L., Kopito, R. R.
  2003; 14 (3): 1268-1278

- **Cystic fibrosis: premature degradation of mutant proteins as a molecular disease mechanism.** *Methods in molecular biology (Clifton, N.J.)*
  Gelman, M. S., Kopito, R. R.
  2003; 232: 27-37

- **Rescuing protein conformation: prospects for pharmacological therapy in cystic fibrosis** *JOURNAL OF CLINICAL INVESTIGATION*
  Gelman, M. S., Kopito, R. R.
  2002; 110 (11): 1591-1597

- **A rhodopsin mutant linked to autosomal dominant retinitis pigmentosa is prone to aggregate and interacts with the ubiquitin proteasome system** *JOURNAL OF BIOLOGICAL CHEMISTRY*

Illing, M. E., Rajan, R. S., Bence, N. F., Kopito, R. R.
2002; 277 (37): 34150-34160

- **Cytoplasmic dynein/dynactin mediates the assembly of aggresomes** *CELL MOTILITY AND THE CYTOSKELETON*
  Johnston, J. A., Illing, M. E., Kopito, R. R.
  2002; 53 (1): 26-38
- **A principal role for the proteasome in endoplasmic reticulum-associated degradation of misfolded intracellular cystic fibrosis transmembrane conductance regulator** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Gelman, M. S., Kannegaard, E. S., Kopito, R. R.
  2002; 277 (14): 11709-11714
- **Cysteine residues in the nucleotide binding domains regulate the conductance state of CFTR channels** *BIOPHYSICAL JOURNAL*
  Harrington, M. A., Kopito, R. R.
  2002; 82 (3): 1278-1292
- **Specificity in intracellular protein aggregation and inclusion body formation** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
  Rajan, R. S., Illing, M. E., Bence, N. F., Kopito, R. R.
  2001; 98 (23): 13060-13065
- **Aggresomes, inclusion bodies and protein aggregation** *TRENDS IN CELL BIOLOGY*
  Kopito, R. R.
  2000; 10 (12): 524-530
- **Formation of high molecular weight complexes of mutant Cu,Zn-superoxide dismutase in a mouse model for familial amyotrophic lateral sclerosis** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
  Johnston, J. A., Dalton, M. J., Gurney, M. E., Kopito, R. R.
  2000; 97 (23): 12571-12576
- **Conformational disease** *NATURE CELL BIOLOGY*
  Kopito, R. R., Ron, D.
  2000; 2 (11): E207-E209
- **Aggresomes and Russell bodies - Symptoms of cellular indigestion?** *EMBO REPORTS*
  Kopito, R. R., Sitia, R.
  2000; 1 (3): 225-231
- **The role of multiubiquitination in dislocation and degradation of the alpha subunit of the T cell antigen receptor** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Yu, H., Kopito, R. R.
  1999; 274 (52): 36852-36858
- **Redox reagents and divalent cations alter the kinetics of cystic fibrosis transmembrane conductance regulator channel gating** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Harrington, M. A., Gunderson, K. L., Kopito, R. R.
  1999; 274 (39): 27536-27544
- **Aggresomes: A cellular response to misfolded proteins**
  Johnston, J. A., Ward, C. L., Kopito, R. R.
  FEDERATION AMER SOC EXP BIOL.1999: A1520–A1520
- **Biosynthesis and degradation of CFTR.** *Physiological reviews*
  Kopito, R. R.
  1999; 79 (1): S167-73
- **Aggresomes: A cellular response to misfolded proteins** *JOURNAL OF CELL BIOLOGY*
  Johnston, J. A., Ward, C. L., Kopito, R. R.
  1998; 143 (7): 1883-1898
- **Aggresomes: A cellular response to misfolded proteins**

Johnson, J. A., Ward, C. W., Kopito, R. R.
AMER SOC CELL BIOLOGY.1998: 260A–260A

- **Indirect requirement for multi-ubiquitination in TCR alpha degradation**
  Yu, H., Kopito, R.
  AMER SOC CELL BIOLOGY.1998: 459A–459A
- **Cytosolic pH regulates G(Cl) through control of phosphorylation states of CFTR** *AMERICAN JOURNAL OF PHYSIOLOGY-CELL PHYSIOLOGY*
  Reddy, M. M., Kopito, R. R., Quinton, P. M.
  1998; 275 (4): C1040-C1047
- **Topology of the region surrounding Glu(681) of human AE1 protein, the erythrocyte anion exchanger** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Tang, X. B., Fujinaga, J., Kopito, R., Casey, J. R.
  1998; 273 (35): 22545-22553
- **Cotranslational ubiquitination of cystic fibrosis transmembrane conductance regulator in vitro** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Sato, S., Ward, C. L., Kopito, R. R.
  1998; 273 (13): 7189-7192
- **A cluster of cytoplasmic histidine residues specifies pH dependence of the AE2 plasma membrane anion exchanger.** *Cell*
  Kobayashi, S., Kopito, R. R.
  1997; 90 (6): following 1159-?
- **Cytosolic degradation of T-cell receptor alpha chains by the proteasome** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Yu, H., Kaung, G., Kobayashi, S., Kopito, R. R.
  1997; 272 (33): 20800-20804
- **Cystic fibrosis transmembrane conductance regulator and adenosine triphosphate - Response** *SCIENCE*
  Reddy, M. M., Quinton, P. M., HAWS, C., Wine, J. J., Grygorczyk, R., Tabcharani, J. A., Hanrahan, J. W., Gunderson, K. L., Kopito, R. R.
  1997; 275 (5304): 1325-1325
- **ER quality control: The cytoplasmic connection** *CELL*
  Kopito, R. R.
  1997; 88 (4): 427-430
- **Determination of topology and pore-lining residues in the transmembrane segment 8 region of AE1, the human erythrocyte anion exchange protein**
  Casey, J. R., Tang, X. B., Kopito, R. R.
  CELL PRESS.1997: TU346–TU346
- **A cluster of cytoplasmic histidine residues specifies pH dependence of the AE2 plasma membrane anion exchanger** *CELL*
  Sekler, I., Kobayashi, S., Kopito, R. R.
  1996; 86 (6): 929-935
- **Immunocytochemical localization of vacuolar H+-ATPase and Cl--HCO3- anion exchanger (Erythrocyte band-3 protein) in avian osteoclasts: Effect of calcium-deficient diet on polar expression of the H+-ATPase pump** *CALCIFIED TISSUE INTERNATIONAL*
  Bastani, B., Ross, F. P., Kopito, R. R., Gluck, S. L.
  1996; 58 (5): 332-336
- **Failure of the cystic fibrosis transmembrane conductance regulator to conduct ATP** *SCIENCE*
  Reddy, M. M., Quinton, P. M., HAWS, C., Wine, J. J., Grygorczyk, R., Tabcharani, J. A., Hanrahan, J. W., Gunderson, K. L., Kopito, R. R.
  1996; 271 (5257): 1876-1879
- **Glycerol reverses the misfolding phenotype of the most common cystic fibrosis mutation** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Sato, S., Ward, C. L., Krouse, M. E., Wine, J. J., Kopito, R. R.
  1996; 271 (2): 635-638
- **DEGRADATION OF CFTR BY THE UBIQUITIN-PROTEASOME PATHWAY** *CELL*
  Ward, C. L., OMURA, S., Kopito, R. R.

1995; 83 (1): 121-127

- **CONFORMATIONAL STATES OF CFTR ASSOCIATED WITH CHANNEL GATING - THE ROLE OF ATP BINDING AND HYDROLYSIS** *CELL*
  Gunderson, K. L., Kopito, R. R.
  1995; 82 (2): 231-239

- **INTRACELLULAR TURNOVER OF CYSTIC-FIBROSIS TRANSMEMBRANE CONDUCTANCE REGULATOR - INEFFICIENT PROCESSING AND RAPID DEGRADATION OF WILD-TYPE AND MUTANT PROTEINS** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Ward, C. L., Kopito, R. R.
  1994; 269 (41): 25710-25718

- **AE3 ANION-EXCHANGER ISOFORMS IN THE VERTEBRATE RETINA - DEVELOPMENTAL REGULATION AND DIFFERENTIAL EXPRESSION IN NEURONS AND GLIA** *JOURNAL OF NEUROSCIENCE*
  Kobayashi, S., Morgans, C. W., Casey, J. R., Kopito, R. R.
  1994; 14 (10): 6266-6279

- **CL-/HCO3- EXCHANGE FUNCTION DIFFERS IN ADULT AND FETAL-RAT HIPPOCAMPAL-NEURONS** *BRAIN RESEARCH*
  RALEYSUSMAN, K. M., Sapolsky, R. M., Kopito, R. R.
  1993; 614 (1-2): 308-314

- **CLONING AND CHARACTERIZATION OF BAND-3, THE HUMAN-ERYTHROCYTE ANION-EXCHANGE PROTEIN (AE1)** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
  Lux, S. E., John, K. M., Kopito, R. R., Lodish, H. F.
  1989; 86 (23): 9089-9093

- **IDENTIFICATION OF A 185-KDA BAND 3-RELATED POLYPEPTIDE IN OXYNTIC CELLS** *AMERICAN JOURNAL OF PHYSIOLOGY*
  Thomas, H. A., Machen, T. E., Smolka, A., Baron, R., Kopito, R. R.
  1989; 257 (3): C537-C544

- **A 115-KD POLYPEPTIDE IMMUNOLOGICALLY RELATED TO ERYTHROCYTE BAND-3 IS PRESENT IN GOLGI MEMBRANES** *SCIENCE*
  Kellokumpu, S., Neff, L., JAMSAKELLOKUMPU, S., Kopito, R., Baron, R.
  1988; 242 (4883): 1308-1311

- **CLONING AND CHARACTERIZATION OF A MURINE BAND 3-RELATED CDNA FROM KIDNEY AND FROM A LYMPHOID-CELL LINE** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Alper, S. L., Kopito, R. R., LIBRESCO, S. M., Lodish, H. F.
  1988; 263 (32): 17092-17099

- **Structure and tissue-specific expression of the mouse anion-exchanger gene in erythroid and renal cells.** *Society of General Physiologists series*
  Kopito, R. R., Andersson, M. M., Herzlinger, D. A., Al-Awqati, Q., Lodish, H. F.
  1988; 43: 151-161

- **A molecular biological approach to the study of anion transport.** *Kidney international. Supplement*
  Alper, S. L., Kopito, R. R., Lodish, H. F.
  1987; 23: S117-33

- **A MOLECULAR BIOLOGICAL APPROACH TO THE STUDY OF ANION TRANSPORT** *KIDNEY INTERNATIONAL*
  Alper, S. L., Kopito, R. R., Lodish, H. F.
  1987; 32: S117-S128

- **MULTIPLE TISSUE-SPECIFIC SITES OF TRANSCRIPTIONAL INITIATION OF THE MOUSE ANION ANTIPORT GENE IN ERYTHROID AND RENAL-CELLS** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
  Kopito, R. R., Andersson, M. A., Lodish, H. F.
  1987; 84 (20): 7149-7153

- **STRUCTURE AND ORGANIZATION OF THE MURINE BAND-3 GENE** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Kopito, R. R., Andersson, M., Lodish, H. F.
  1987; 262 (17): 8035-8040

- **RADIOENZYMATIC ASSAY OF PLASMA MEVALONATE** *METHODS IN ENZYMOLOGY*

Parker, T. S., Kopito, R. R., Brunengraber, H.
1985; 110: 58-71

- **STRUCTURE OF THE MURINE ANION-EXCHANGE PROTEIN** *JOURNAL OF CELLULAR BIOCHEMISTRY*
  Kopito, R. R., Lodish, H. F.
  1985; 29 (1): 1-17

- **PRIMARY STRUCTURE AND TRANSMEMBRANE ORIENTATION OF THE MURINE ANION-EXCHANGE PROTEIN** *NATURE*
  Kopito, R. R., Lodish, H. F.
  1985; 316 (6025): 234-238

- **THE SHUNT PATHWAY OF MEVALONATE METABOLISM IN THE ISOLATED PERFUSED RAT-KIDNEY** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  Kopito, R. R., Murray, D. M., STORY, D. L., Brunengraber, H.
  1984; 259 (1): 372-377

- **THE SHUNT PATHWAY OF MEVALONATE METABOLISM IN THE ISOLATED PERFUSED-RAT-LIVER** *JOURNAL OF BIOLOGICAL CHEMISTRY*
  WEINSTOCK, S. B., Kopito, R. R., Endemann, G., Tomera, J. F., Marinier, E., Murray, D. M., Brunengraber, H.
  1984; 259 (14): 8939-8944

- **ASSESSMENT OF THE FLUX OF MITOCHONDRIAL ACETYL-COA IN LIVER AND KIDNEY BY USING THE DIFFERENTIAL PRODUCTION OF (CO2)-C-14 FROM TRACERS OF (1-C-14)-LABELED AND (2-C-14)-LABELED 4-METHYL-2-OXOVALERATE** *BIOCHEMICAL JOURNAL*
  Tomera, J. F., Kopito, R. R., Brunengraber, H.
  1983; 210 (1): 265-268

- **METABOLISM OF PLASMA MEVALONATE IN RATS AND HUMANS** *JOURNAL OF LIPID RESEARCH*
  Kopito, R. R., WEINSTOCK, S. B., Freed, L. E., Murray, D. M., Brunengraber, H.
  1982; 23 (4): 577-583

- **URINARY CLEARANCE AND METABOLISM OF MEVALONATE BY THE ISOLATED PERFUSED RAT-KIDNEY** *JOURNAL OF LIPID RESEARCH*
  Brunengraber, H., WEINSTOCK, S. B., STORY, D. L., Kopito, R. R.
  1981; 22 (6): 916-920

- **(R)-MEVALONATE EXCRETION IN HUMAN AND RAT URINES** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA-BIOLOGICAL SCIENCES*
  Kopito, R. R., Brunengraber, H.
  1980; 77 (10): 5738-5740

- **SIALOPROTEINEMIA - LACK OF CORRELATION WITH INHIBITION OF INVITRO LYMPHOBLASTOSIS INDUCED BY PHYTOHEMAGGLUTININ OR ALLOANTIGEN** *CLINICAL AND EXPERIMENTAL IMMUNOLOGY*
  Gray, B. N., Kopito, R. R., Anderson, L. L., BARALT, O. L., CONNERY, C. K., Watkins, E.
  1976; 25 (2): 227-233