Stanford

Ron Kopito

Professor of Biology

CONTACT INFORMATION

- **Alternate Contact**

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Bio

ACADEMIC APPOINTMENTS

- Professor, Biology
- Member, Bio-X
- 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

2018-19

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

2017-18

- Advanced Cell Biology: BIO 214, BIOC 224, MCP 221 (Win)
- Cystic fibrosis: from medical conundrum to precision medicine success story: BIO 25Q (Spr)
- Molecular Mechanisms of Neurodegenerative Disease: BIO 267, GENE 267, NENS 267 (Win)

2016-17

- Advanced Cell Biology: BIO 214, BIOC 224, MCP 221 (Win)
- Understanding Neurodegenerative Disease: OSPPARIS 87 (Win)

2015-16

- Advanced Cell Biology: BIO 214, BIOC 224, MCP 221 (Win)
- Molecular Mechanisms of Neurodegenerative Disease: BIO 267, GENE 267, NENS 267 (Win)
- The Molecular Basis of Genetic Disease: BIO 25Q (Spr)

STANFORD ADVISEES

Doctoral Dissertation Reader (AC)

Abel Ferrel, Naomi Genuth, Darius Johnston, Pin-Joe Ko, Laura Persson

Postdoctoral Faculty Sponsor

Paul Da Rosa, Celeste Riepe

GRADUATE AND FELLOWSHIP PROGRAM AFFILIATIONS

- Biochemistry (Phd Program)
- Biology (School of Humanities and Sciences) (Phd Program)
- Biophysics (Phd Program)
- Neurosciences (Phd Program)

Publications

PUBLICATIONS

- **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

- **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

- **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

- **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. P., Leto, D. E., Shaler, T. A., Kopito, R. R.
  2018; 70 (3): 516-+

- **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)

- **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

- **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

- **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-U176
- **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

- **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

- **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

- **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.
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- **Misfolded proteins partition between two distinct quality control compartments** *NATURE*
  Kaganovich, D., Kopito, R., Frydman, J.
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- **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

- **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.
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- **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

- **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.
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- **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

- **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

- **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

- **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

- **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.
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- **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

- **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

- **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.
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- **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.
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- **Recognition of a single transmembrane degron by sequential quality control checkpoints** *MOLECULAR BIOLOGY OF THE CELL*
  Fayadat, L., Kopito, R. R.
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- **Cystic fibrosis: premature degradation of mutant proteins as a molecular disease mechanism.** *Methods in molecular biology (Clifton, N.J.)*
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- **Rescuing protein conformation: prospects for pharmacological therapy in cystic fibrosis** *JOURNAL OF CLINICAL INVESTIGATION*
  Gelman, M. S., Kopito, R. R.
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- **A rhodopsin mutant linked to autosomal dominant retinitis pigmentosa is prone to aggregate and interacts with the ubiquitin proteasome system** *JOURNAL OF BIOLOGICAL CHEMISTRY*
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  Johnston, J. A., Illing, M. E., Kopito, R. R.
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- **A principal role for the proteasome in endoplasmic reticulum-associated degradation of misfolded intracellular cystic fibrosis transmembrane conductance regulator** *JOURNAL OF BIOLOGICAL CHEMISTRY*
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- **Cysteine residues in the nucleotide binding domains regulate the conductance state of CFTR channels** *BIOPHYSICAL JOURNAL*
  Harrington, M. A., Kopito, R. R.
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- **Specificity in intracellular protein aggregation and inclusion body formation** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
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- **Impairment of the ubiquitin-proteasome system by protein aggregation** *SCIENCE*
  Bence, N. F., Sampat, R. M., Kopito, R. R.
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- **Aggresomes, inclusion bodies and protein aggregation** *TRENDS IN CELL BIOLOGY*
  Kopito, R. R.
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- **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*
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  Kopito, R. R., Ron, D.

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- **Aggresomes and Russell bodies - Symptoms of cellular indigestion?** *EMBO REPORTS*
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- **Redox reagents and divalent cations alter the kinetics of cystic fibrosis transmembrane conductance regulator channel gating** *JOURNAL OF BIOLOGICAL CHEMISTRY*
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- **Aggresomes: A cellular response to misfolded proteins**
  Johnson, J. A., Ward, C. W., Kopito, R. R.
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  Yu, H., Kopito, R.
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  Reddy, M. M., Kopito, R. R., Quinton, P. M.
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  Yu, H., Kaung, G., Kobayashi, S., Kopito, R. R.
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  Sekler, I., Kobayashi, S., Kopito, R. R.
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