Ron Rieger Kopito

Professor of Biology. kopito@stanford.edu. 650.723.7581 Born:
December 21, 1954
U.S. Citizen

Stanford University Department
of Biology
327 Campus Drive Stanford, California 94305

Professional Experience

Stanford University *Professor,*
Department of Biology Stanford, *1996 – present*
California 94305

Associate Professor, Department of Biology *1992 - 1996*
Stanford, California 94305

Assistant Professor, Department of Biology *1987 - 1992*
Stanford, California 94305

MIT/Whitehead Institute
Postdoctoral Fellow *1982-1986*
Mentor: Harvey Lodish
Cambridge, Massachusetts 02142

Education

Massachusetts Institute of Technology *June 1982*
Ph.D. Nutritional Biochemistry and Metabolism
Mentor: Henri Brunengraber
Cambridge, Massachusetts 02139

Bowdoin College *June 1976*
A.B. Biochemistry
Brunswick, Maine 04011

Honors and Awards

1982	NIH Postdoctoral Fellowship
1985	Lucille P. Markey Scholar in Biomedical Science
1989	National Science Foundation Presidential Young Investigator
1989	March of Dimes Basil O'Connor Starter Scholar Research Award

1993 American Heart Association Established Investigatorship Award 2018
Fellow of the American Society for Cell Biology

Conferences Organized

1995 Chair, Gordon Research Conference: "Mechanisms of Membrane Transport"
2002 Chair, FASEB Summer conference: "Amyloids and other abnormal protein folding processes"
2002 Co-chair, Society of General Physiologists Conference: "Assembly and trafficking of transporters"
2011 - 2015 Co-organizer, Cold Spring Harbor Conference: "The Ubiquitin Family"

Reviewing and Editorial

1988 - Ad hoc reviewer, NIH
1989 – 1999 Editorial Board, *J. Biol. Chem.*
1995 - Editorial Board, *J. Cell Sci.*
2005 - Editorial Board, J. Membrane Biol.
2006 - Editorial Board, *Autophagy*
2011 – 2016 Member, MBPP study section NIH
2011 - Editorial Board, *Journal of Huntington's Disease*

Professional Societies

1982 – present American Society for Cell Biology
1988 – present Society of General Physiologists
1990 – present Biophysical Society
1991 – present American Society for Biochemistry and Molecular Biology

Patents

2010 "Composition and Methods for High Throughput Screening of Pharmacological Chaperones" - Ron R. Kopito and Wei Zhang: US Patent 7,790,364 B2

Departmental and University Service

2020 – present Chair, Biology Department Safety Committee
2024- present, Member, University Health and Safety Committee
2000- present, Member, Advisory Board, Protein and Nucleic Acid (PAN) facility

Publications

1. Gray, B.N., R.R. Kopito, L.L. Anderson, O.L. Baralt, C.K. Connery and E. Watkins, Jr. (1976) Sialoproteinaemia: lack of correlation with inhibition of in vitro lymphoblastosis induced by phytohaemagglutinin or alloantigen. *Clin Exp Immunol*, 25(2), 227-33.
2. Kopito, R.R. and H. Brunengraber (1980) (R)-mevalonate excretion in human and rat urines. *Proc Natl Acad Sci U S A*, 77(10), 5738-40.
3. Brunengraber, H., S.B. Weinstock, D.L. Story and R.R. Kopito (1981) Urinary clearance and metabolism of mevalonate by the isolated perfused rat kidney. *J Lipid Res*, 22(6), 916-20.

4. Kopito, R.R., S.B. Weinstock, L.E. Freed, D.M. Murray and H. Brunengraber (1982) Metabolism of plasma mevalonate in rats and humans. *J Lipid Res*, 23(4), 577-83.
5. Tomera, J.F., R.R. Kopito and H. Brunengraber (1983) Assessment of the flux of mitochondrial acetyl-CoA in liver and kidney by using the differential production of 14CO2 from tracers of (1-14C)- and (2-14C)-labeled 4-methyl-2-oxovalerate. *Biochem J*, 210(1), 265-8.
6. Kopito, R.R., D.M. Murray, D.L. Story and H. Brunengraber (1984) The shunt pathway of mevalonate metabolism in the isolated perfused rat kidney. *J Biol Chem*, 259(1), 372-7.
7. Weinstock, S.B., R.R. Kopito, G. Endemann, J.F. Tomera, E. Marinier, D.M. Murray and H. Brunengraber (1984) The shunt pathway of mevalonate metabolism in the isolated perfused rat liver. *J Biol Chem*, 259(14), 8939-44.
8. Kopito, R.R. and H.F. Lodish (1985) Primary structure and transmembrane orientation of the murine anion exchange protein. *Nature*, 316(6025), 234-8.
9. Kopito, R.R. and H.F. Lodish (1985) Structure of the murine anion exchange protein. *J Cell Biochem*, 29(1), 1-17.
10. Parker, T.S., R.R. Kopito and H. Brunengraber (1985) Radioenzymatic assay of plasma mevalonate. *Methods Enzymol*, 110, 58-71.
11. Alper, S.L., R.R. Kopito and H.F. Lodish (1987) A molecular biological approach to the study of anion transport. *Kidney Int Suppl*, 23, S117-33.
12. Kopito, R.R., M.A. Andersson and H.F. Lodish (1987) Multiple tissue-specific sites of transcriptional initiation of the mouse anion antiport gene in erythroid and renal cells. *Proc Natl Acad Sci U S A*, 84(20), 7149-53.
13. Kopito, R.R., M. Andersson and H.F. Lodish (1987) Structure and organization of the murine band 3 gene. *J Biol Chem*, 262(17), 8035-40.
14. Alper, S.L., R.R. Kopito, S.M. Libresco and H.F. Lodish (1988) Cloning and characterization of a murine band 3-related cDNA from kidney and from a lymphoid cell line. *J Biol Chem*, 263(32), 17092-9.
15. Kellokumpu, S., L. Neff, S. Jamsa-4, R. Kopito and R. Baron (1988) A 115-kD polypeptide immunologically related to erythrocyte band 3 is present in Golgi membranes. *Science*, 242(4883), 1308-11.
16. Kopito, R.R., M.M. Andersson, D.A. Herzlinger, Q. al-Awqati and H.F. Lodish (1988) Structure and tissue-specific expression of the mouse anion-exchanger gene in erythroid and renal cells. *Soc Gen Physiol Ser*, 43, 151-61.
17. Kopito, R.R., B.S. Lee, D.M. Simmons, A.E. Lindsey, C.W. Morgans and K. Schneider (1989) Regulation of intracellular pH by a neuronal homolog of the erythrocyte anion exchanger. *Cell*, 59(5), 927-37.
18. Lux, S.E., K.M. John, R.R. Kopito and H.F. Lodish (1989) Cloning and characterization of band 3, the human erythrocyte anion-exchange protein (AE1). *Proc Natl Acad Sci U S A*, 86(23), 9089-93.
19. Stewart, E.A., R. Kopito and A.M. Bowcock (1989) A PstI polymorphism for the human erythrocyte surface protein band 3 (EPB3) demonstrates close linkage of EPB3 to the nerve growth factor receptor. *Genomics*, 5(3), 633-5.
20. Thomas, H.A., T.E. Machen, A. Smolka, R. Baron and R.R. Kopito (1989) Identification of a 185-kDa band 3-related polypeptide in oxyntic cells. *Am J Physiol*, 257(3 Pt 1), C537-44.
21. Kopito, R.R. (1990) Molecular biology of the anion exchanger gene family. *Int Rev Cytol*, 123, 177-99.

22. Lindsey, A.E., K. Schneider, D.M. Simmons, R. Baron, B.S. Lee and R.R. Kopito (1990) Functional expression and subcellular localization of an anion exchanger cloned from choroid plexus. *Proc Natl Acad Sci U S A*, 87(14), 5278-82.
23. Lee, B.S., R.B. Gunn and R.R. Kopito (1991) Functional differences among nonerythroid anion exchangers expressed in a transfected human cell line. *J Biol Chem*, 266(18), 11448-54.
24. Raley-Susman, K.M., E.J. Cragoe, Jr., R.M. Sapolsky and R.R. Kopito (1991) Regulation of intracellular pH in cultured hippocampal neurons by an amiloride-insensitive Na+/H+ exchanger. *J Biol Chem*, 266(5), 2739-45.
25. Ward, C.L., M.E. Krouse, D.C. Gruenert, R.R. Kopito and J.J. Wine (1991) Cystic fibrosis gene expression is not correlated with rectifying Cl- channels. *Proc Natl Acad Sci U S A*, 88(12), 5277-81.
26. Morgans, C.W. and R.R. Kopito (1993) Generation of truncated brain AE3 isoforms by alternate mRNA processing. *J Cell Sci*, 106 (Pt 4), 1275-82.
27. Morgans, C.W. and R.R. Kopito (1993) Association of the brain anion exchanger, AE3, with the repeat domain of ankyrin. *J Cell Sci*, 105 (Pt 4), 1137-42.
28. Raley-Susman, K.M., R.M. Sapolsky and R.R. Kopito (1993) Cl-/HCO3- exchange function differs in adult and fetal rat hippocampal neurons. *Brain Res*, 614(1-2), 308-14.
29. Ruetz, S., A.E. Lindsey, C.L. Ward and R.R. Kopito (1993) Functional activation of plasma membrane anion exchangers occurs in a pre-Golgi compartment. *J Cell Biol*, 121(1), 37-48.
30. Ruetz, S., A.E. Lindsey and R.R. Kopito (1993) Function and biosynthesis of erythroid and nonerythroid anion exchangers. *Soc Gen Physiol Ser*, 48, 193-200.
31. Ding, Y., J.R. Casey and R.R. Kopito (1994) The major kidney AE1 isoform does not bind ankyrin (Ank1) in vitro. An essential role for the 79 NH2-terminal amino acid residues of band 3. *J Biol Chem*, 269(51), 32201-8.
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33. Kobayashi, S., C.W. Morgans, J.R. Casey and R.R. Kopito (1994) AE3 anion exchanger isoforms in the vertebrate retina: developmental regulation and differential expression in neurons and glia. *J Neurosci*, 14(10), 6266-79.
34. Ward, C.L. and R.R. Kopito (1994) Intracellular turnover of cystic fibrosis transmembrane conductance regulator. Inefficient processing and rapid degradation of wild-type and mutant proteins. *J Biol Chem*, 269(41), 25710-8.
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41. Bastani, B., F.P. Ross, R.R. Kopito and S.L. Gluck (1996) 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. *Calcif Tissue Int*, 58(5), 332-6.
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44. Sato, S., C.L. Ward, M.E. Krouse, J.J. Wine and R.R. Kopito (1996) Glycerol reverses the misfolding phenotype of the most common cystic fibrosis mutation. *J Biol Chem*, 271(2), 635-8.
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47. Kobayashi, S. and R.R. Kopito (1997) A cluster of cytoplasmic histidine residues specifies pH dependence of the AE2 plasma membrane anion exchanger. *Cell*, 90(6), following 1159.
48. Kopito, R.R. (1997) ER quality control: the cytoplasmic connection. *Cell*, 88(4), 427-30.
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50. Zeng, W., M.G. Lee, M. Yan, J. Diaz, I. Benjamin, C.R. Marino, R. Kopito, S. Freedman, C. Cotton, S. Muallem and P. Thomas (1997) Immuno and functional characterization of CFTR in submandibular and pancreatic acinar and duct cells. *Am J Physiol*, 273(2 Pt 1), C442-55.
51. Johnston, J.A., C.L. Ward and R.R. Kopito (1998) Aggresomes: a cellular response to misfolded proteins. *J Cell Biol*, 143(7), 1883-98.
52. Kopito, R.R., (1998) *Ubiquitination of integral membrane proteins and proteins in the secretory pathway*, In *Ubiquitin and the Biology of the Cell*, J.-M. Peters, Harris, J.R. and Finley, D.,ed. Plenum, New York. pp. 389-407.
53. Reddy, M.M., R.R. Kopito and P.M. Quinton (1998) Cytosolic pH regulates GCl through control of phosphorylation states of CFTR. *Am J Physiol*, 275(4 Pt 1), C1040-7.
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57. Kopito, R.R. (1999) Biosynthesis and degradation of CFTR. *Physiol Rev*, 79(1 Suppl), S167-73.
58. Yu, H. and R.R. Kopito (1999) The role of multiubiquitination in dislocation and degradation of the alpha subunit of the T cell antigen receptor. *J Biol Chem*, 274(52), 36852-8.
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61. Kopito, R.R. (2000) Aggresomes, inclusion bodies and protein aggregation. *Trends Cell Biol*, 10(12), 524-30.
62. Kopito, R.R. and D. Ron (2000) Conformational disease. *Nat Cell Biol*, 2(11), E207-9.
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64. Rajan, R.S., M.E. Illing, N.F. Bence and R.R. Kopito (2001) Specificity in intracellular protein aggregation and inclusion body formation. *Proc Natl Acad Sci U S A*, 98(23), 13060-5.
65. Gelman, M.S., E.S. Kannegaard and R.R. Kopito (2002) A principal role for the proteasome in endoplasmic reticulum-associated degradation of misfolded intracellular cystic fibrosis transmembrane conductance regulator. *J Biol Chem*, 277(14), 11709-14.
66. Harrington, M.A. and R.R. Kopito (2002) Cysteine residues in the nucleotide binding domains regulate the conductance state of CFTR channels. *Biophys J*, 82(3), 1278-92.
67. Illing, M.E., R.S. Rajan, N.F. Bence and R.R. Kopito (2002) A rhodopsin mutant linked to autosomal dominant retinitis pigmentosa is prone to aggregate and interacts with the ubiquitin proteasome system. J Biol Chem, 277(37), 34150-60.
68. Johnston, J.A., M.E. Illing and R.R. Kopito (2002) Cytoplasmic dynein/dynactin mediates the assembly of aggresomes. Cell Motil Cytoskeleton, 53(1), 26-38.
69. Lenk, U., H. Yu, J. Walter, M.S. Gelman, E. Hartmann, R.R. Kopito and T. Sommer (2002) A role for mammalian Ubc6 homologues in ER-associated protein degradation. J Cell Sci, 115(Pt 14), 3007-14.
70. M.S. Gelman and R.R. Kopito (2002) Rescuing protein conformation: prospects for pharmacological therapy in cystic fibrosis. J. Clin. Invest.110:1591-1597.
71. Fayadat L and R.R. Kopito (2003) Recognition of a Single Transmembrane Degron by Sequential Quality Control Checkpoints. Molecular Biology of the Cell, Vol.14, 1268-1278, March 2003
72. Yechiel Elkabetz, Anat Kerem, Lilach Tencer, Dorit Winitz, R.R. Kopito and Shoshana Bar-Nun (2003). Immunoglobulin Light Chains Dictate Vesicular Transport-dependent and –independent Routes for IgM Degradation by the Ubiquitin-Proteasome Pathway. The Journal of Biological Chemistry. Vol. 278, No. 21,18922-18929.
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75. Rajan, R.S. and R.R. Kopito, (2005) Suppression of wild-type rhodopsin maturation by mutants linked to autosomal dominant retinitis pigmentosa. J Biol Chem. 280:1284-1291.
76. Gilchrist C.A., D.A. Gray, A. Stieber, N.K. Gonatas, R.R. Kopito. (2005) Effect of ubiquitin expression on neuropathogenesis in a mouse model of familial amyotrophic lateral sclerosis. Neuropathol Appl Neurobiol. 31:20-33.
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90. Ryu, K.Y., 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., R.R. Kopito. The mouse polyubiquitin gene Ubb is essential for meiotic progression. *Mol. Cell Biol.*, 2008, 28(3):1136-46. PMCID: PMC2223379
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