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BORN: December 21, 1954
U.S. Citizen

Professional Experience

1996 -
STANFORD UNIVERSITY
Professor of Biological Sciences
Department of Biology
Stanford, CA 94305-5020

1992 - 1996
STANFORD UNIVERSITY
Associate Professor
Department of Biology
Stanford, California 94305-5020

1987 - 1992
STANFORD UNIVERSITY
Assistant Professor
Department of Biology
Stanford, California 94305-5020

1982-1986
MASSACHUSETTS INSTITUTE OF TECHNOLOGY
and WHITEHEAD INSTITUTE FOR BIOMEDICAL RESEARCH
Postdoctoral Fellow
Mentor: Harvey Lodish
Cambridge, Massachusetts 02142

Education

Ph.D. MASSACHUSETTS INSTITUTE OF TECHNOLOGY
Nutritional Biochemistry and Metabolism
Mentor: Henri Brunengraber
Cambridge, Massachusetts 02139
June, 1982

A.B., cum laude BOWDOIN COLLEGE
Biochemistry
Brunswick, Maine 04011
June, 1976

Honors and Awards

1993-1998 American Heart Association Established Investigatorship Award
1989-1994 National Science Foundation Presidential Young Investigator

1989-1992 March of Dimes Basil O'Connor Starter Scholar Research Award
1985-1991 Lucille P. Markey Scholar in Biomedical Science
1982-1985 NIH Postdoctoral Fellowship

Conferences Organized

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

Reviewing and Editorial

1995- Editorial Board, *J. Cell Sci.*
1989-1999 Editorial Board, *J. Biol. Chem.*
2006- Editorial Board, *Autophagy*
2005- Editorial Board, *J. Membrane Biol.*
1988- Ad hoc reviewer, NIH
2011- Member, MBPP study section NIH (beginning 9/11)
2011- Editorial Board, Journal of Huntington's Disease

Professional Societies

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

Patents

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

Departmental and University Service

2007- 2011 Chair, Graduate Admissions Committee
2007- 2011 Representative to Committee on Graduate Education and Policy (CGAP)
2011- 2012 Chair, Biology Department Seminar Committee

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 isolataed 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)-labelled 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-Kellokumpu, 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.
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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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a role for ATP hydrolysis in cystic fibrosis transmembrane regulator channel gating. *J Biol Chem*, **269**(30), 19349-53.
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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46. Ward, C.L.a.K., R.R., (1996) *Biosynthesis and degradation of CFTR*, In *Cystic Fibrosis*Current Topics*, J.A. In Dodge, Brock, J.H. and Widdicombe, J.H. (eds),ed. John wiley & Sons Ltd., New York. pp. 107-115.
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.
49. Yu, H., G. Kaung, S. Kobayashi and R.R. Kopito (1997) Cytosolic degradation of T-cell receptor alpha chains by the proteasome. *J Biol Chem*, **272**(33), 20800-4.
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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.
59. Johnston, J.A., M.J. Dalton, M.E. Gurney and R.R. Kopito (2000) Formation of high molecular weight complexes of mutant Cu, Zn-superoxide dismutase in a mouse model for familial amyotrophic lateral sclerosis. *Proc Natl Acad Sci U S A*, **97**(23), 12571-6.

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63. Bence, N.F., R.M. Sampat and R.R. Kopito (2001) Impairment of the ubiquitin-proteasome system by protein aggregation. *Science*, **292**(5521), 1552-5.
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.
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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.
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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.
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83. Ryu K.Y., R.T. Baker, R.R. Kopito. (2006) Ubiquitin-specific protease 2 as a tool for quantification of total ubiquitin levels in biological specimens. Anal. Biochem. 353(1):153-155.
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86. Bruns, C.K. and R.R. Kopito. (2007) Impaired posttranslational folding of familial ALS-linked Cu, Zn superoxide dismutase mutants. *EMBO J.* 26(3):855-866. PMCID: PMC2892477
87. Ryu, K.Y., Maehr, R., Gilchrist, C.A., Long, M.A., Bouley D.M., Mueller, B., Ploegh, H.L., R.R. Kopito. The mouse polyubiquitin gene UbC is essential for fetal liver development, cell-cycle progression and stress tolerance. *EMBO J.,* 2007, 26(11):2693-2706. PMCID: PMC1888680

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