Richard A Steet

List of Publications by Year
in descending order

26 papers	890 citations	516710 16 h-index	552781 26 g-index
27 all docs	27 docs citations	27 times ranked	1301 citing authors

#	Article	IF	Citations
1	The iminosugar isofagomine increases the activity of N370S mutant acid beta-glucosidase in Gaucher fibroblasts by several mechanisms. Proceedings of the National Academy of Sciences of the United States of America, 2006, 103, 13813-13818.	7.1	198
2	One-Step Selective Exoenzymatic Labeling (SEEL) Strategy for the Biotinylation and Identification of Glycoproteins of Living Cells. Journal of the American Chemical Society, 2016, 138, 11575-11582.	13.7	81
3	COG-7-deficient Human Fibroblasts Exhibit Altered Recycling of Golgi Proteins. Molecular Biology of the Cell, 2006, 17, 2312-2321.	2.1	61
4	Selective action of the iminosugar isofagomine, a pharmacological chaperone for mutant forms of acid-Î²-glucosidase. Biochemical Pharmacology, 2007, 73, 1376-1383.	4.4	48
5	Selective Exo-Enzymatic Labeling Detects Increased Cell Surface Sialoglycoprotein Expression upon Megakaryocytic Differentiation. Journal of Biological Chemistry, 2016, 291, 3982-3989.	3.4	45
6	A zebrafish model of PMM2-CDG reveals altered neurogenesis and a substrate-accumulation mechanism for N-linked glycosylation deficiency. Molecular Biology of the Cell, 2012, 23, 4175-4187.	2.1	44
7	Analysis of Mucolipidosis II/III GNPTAB Missense Mutations Identifies Domains of UDP-GlcNAc:lysosomal Enzyme GlcNAc-1-phosphotransferase Involved in Catalytic Function and Lysosomal Enzyme Recognition. Journal of Biological Chemistry, 2015, 290, 3045-3056.	3.4	42
8	Altered Chondrocyte Differentiation and Extracellular Matrix Homeostasis in a Zebrafish Model for Mucolipidosis II. American Journal of Pathology, 2009, 175, 2063-2075.	3.8	38
9	A splicing mutation in the ?/? GlcNAc-1-phosphotransferase gene results in an adult onset form of mucolipidosis III associated with sensory neuropathy and cardiomyopathy. American Journal of Medical Genetics, Part A, 2005, 132A, 369-375.	1.2	36
10	Excessive activity of cathepsin K is associated with cartilage defects in a zebrafish model of mucolipidosis II. DMM Disease Models and Mechanisms, 2012, 5, 177-190.	2.4	36
11	Cyclopropenone-caged Sondheimer diyne (dibenzo[a,e]cyclooctadiyne): a photoactivatable linchpin for efficient SPAAC crosslinking. Chemical Communications, 2016, 52, 553-556.	4.1	35
12	Identification of the Minimal Lysosomal Enzyme Recognition Domain in Cathepsin D. Journal of Biological Chemistry, 2005, 280, 33318-33323.	3.4	34
13	De Novo Variants in LMNB1 Cause Pronounced Syndromic Microcephaly and Disruption of Nuclear Envelope Integrity. American Journal of Human Genetics, 2020, 107, 753-762.	6.2	30
14	Extensive Mannose Phosphorylation on Leukemia Inhibitory Factor (LIF) Controls Its Extracellular Levels by Multiple Mechanisms. Journal of Biological Chemistry, 2011, 286, 24855-24864.	3.4	23
15	Cathepsin-Mediated Alterations in TGFÃŸ-Related Signaling Underlie Disrupted Cartilage and Bone Maturation Associated With Impaired Lysosomal Targeting. Journal of Bone and Mineral Research, 2016, 31, 535-548.	2.8	18
16	TGF-ÃŸ Regulates Cathepsin Activation during Normal and Pathogenic Development. Cell Reports, 2018, 22, 2964-2977.	6.4	17
17	The translocon-associated protein (TRAP) complex regulates quality control of N-linked glycosylation during ER stress. Science Advances, 2021, 7, .	10.3	17
18	Lysosomal cholesterol accumulation contributes to the movement phenotypes associated with NUS1 haploinsufficiency. Genetics in Medicine, 2021, 23, 1305-1314.	2.4	17

#	Article	IF	Citations
19	Functional analysis of a novel mutation in the TIMM8A gene that causes deafnessâ€dystoniaâ€optic neuronopathy syndrome. Molecular Genetics & Genomic Medicine, 2020, 8, e1121.	1.2	16
20	Selective inhibition of N-linked glycosylation impairs receptor tyrosine kinase processing. DMM Disease Models and Mechanisms, 2019, 12, .	2.4	14
21	The Role of Hematopoietic Cell Transplant in the Glycoprotein Diseases. Cells, 2020, 9, 1411.	4.1	14
22	Latency-associated Peptide of Transforming Growth Factor-Î²1 Is Not Subject to Physiological Mannose Phosphorylation. Journal of Biological Chemistry, 2012, 287, 7526-7534.	3.4	10
23	Phenylbutyrate modulates polyamine acetylase and ameliorates Snyder-Robinson syndrome in a Drosophila model and patient cells. JCI Insight, 2022, 7, .	5.0	7
24	Upregulation of Sortilin, a Lysosomal Sorting Receptor, Corresponds with Reduced Bioavailability of Latent TGFÎ² in Mucolipidosis II Cells. Biomolecules, 2020, 10, 670.	4.0	4
25	Protease-dependent defects in N-cadherin processing drive PMM2-CDG pathogenesis. JCI Insight, 2021, 6, .	5.0	3
26	A Biochemical Platform to Define the Relative Specific Activity of IDUA Variants Identified by Newborn Screening. International Journal of Neonatal Screening, 2020, 6, 88.	3.2	1