

# Richard A Steet

## List of Publications by Year in descending order

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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	Phenylbutyrate modulates polyamine acetylase and ameliorates Snyder-Robinson syndrome in a <i>Drosophila</i> model and patient cells. <i>JCI Insight</i> , 2022, 7, .	5.0	7
2	The translocon-associated protein (TRAP) complex regulates quality control of N-linked glycosylation during ER stress. <i>Science Advances</i> , 2021, 7, .	10.3	17
3	Lysosomal cholesterol accumulation contributes to the movement phenotypes associated with NUS1 haploinsufficiency. <i>Genetics in Medicine</i> , 2021, 23, 1305-1314.	2.4	17
4	Protease-dependent defects in N-cadherin processing drive PMM2-CDG pathogenesis. <i>JCI Insight</i> , 2021, 6, .	5.0	3
5	Functional analysis of a novel mutation in the <i>TIMM8A</i> gene that causes deafness&#x2013;dystonia&#x2013;optic neuropathy syndrome. <i>Molecular Genetics &amp; Genomic Medicine</i> , 2020, 8, e1121.	1.2	16
6	A Biochemical Platform to Define the Relative Specific Activity of IDUA Variants Identified by Newborn Screening. <i>International Journal of Neonatal Screening</i> , 2020, 6, 88.	3.2	1
7	De Novo Variants in LMNB1 Cause Pronounced Syndromic Microcephaly and Disruption of Nuclear Envelope Integrity. <i>American Journal of Human Genetics</i> , 2020, 107, 753-762.	6.2	30
8	Upregulation of Sortilin, a Lysosomal Sorting Receptor, Corresponds with Reduced Bioavailability of Latent TGF $\beta$ <sup>2</sup> in Mucopolidosis II Cells. <i>Biomolecules</i> , 2020, 10, 670.	4.0	4
9	The Role of Hematopoietic Cell Transplant in the Glycoprotein Diseases. <i>Cells</i> , 2020, 9, 1411.	4.1	14
10	Selective inhibition of N-linked glycosylation impairs receptor tyrosine kinase processing. <i>DMM Disease Models and Mechanisms</i> , 2019, 12, .	2.4	14
11	TGF- $\beta$ Regulates Cathepsin Activation during Normal and Pathogenic Development. <i>Cell Reports</i> , 2018, 22, 2964-2977.	6.4	17
12	Cathepsin-Mediated Alterations in TGF- $\beta$ -Related Signaling Underlie Disrupted Cartilage and Bone Maturation Associated With Impaired Lysosomal Targeting. <i>Journal of Bone and Mineral Research</i> , 2016, 31, 535-548.	2.8	18
13	One-Step Selective Exoenzymatic Labeling (SEEL) Strategy for the Biotinylation and Identification of Glycoproteins of Living Cells. <i>Journal of the American Chemical Society</i> , 2016, 138, 11575-11582.	13.7	81
14	Selective Exo-Enzymatic Labeling Detects Increased Cell Surface Sialoglycoprotein Expression upon Megakaryocytic Differentiation. <i>Journal of Biological Chemistry</i> , 2016, 291, 3982-3989.	3.4	45
15	Cyclopropanone-caged Sondheimer diyne (dibenzo[a,e]cyclooctadiyne): a photoactivatable linchpin for efficient SPAAC crosslinking. <i>Chemical Communications</i> , 2016, 52, 553-556.	4.1	35
16	Analysis of Mucopolidosis II/III GNPTAB Missense Mutations Identifies Domains of UDP-GlcNAc:lysosomal Enzyme GlcNAc-1-phosphotransferase Involved in Catalytic Function and Lysosomal Enzyme Recognition. <i>Journal of Biological Chemistry</i> , 2015, 290, 3045-3056.	3.4	42
17	A zebrafish model of PMM2-CDG reveals altered neurogenesis and a substrate-accumulation mechanism for N-linked glycosylation deficiency. <i>Molecular Biology of the Cell</i> , 2012, 23, 4175-4187.	2.1	44
18	Latency-associated Peptide of Transforming Growth Factor- $\beta$ <sup>1</sup> Is Not Subject to Physiological Mannose Phosphorylation. <i>Journal of Biological Chemistry</i> , 2012, 287, 7526-7534.	3.4	10

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19	Excessive activity of cathepsin K is associated with cartilage defects in a zebrafish model of mucopolipidosis II. <i>DMM Disease Models and Mechanisms</i> , 2012, 5, 177-190.	2.4	36
20	Extensive Mannose Phosphorylation on Leukemia Inhibitory Factor (LIF) Controls Its Extracellular Levels by Multiple Mechanisms. <i>Journal of Biological Chemistry</i> , 2011, 286, 24855-24864.	3.4	23
21	Altered Chondrocyte Differentiation and Extracellular Matrix Homeostasis in a Zebrafish Model for Mucopolipidosis II. <i>American Journal of Pathology</i> , 2009, 175, 2063-2075.	3.8	38
22	Selective action of the iminosugar isofagomine, a pharmacological chaperone for mutant forms of acid- $\beta$ -glucosidase. <i>Biochemical Pharmacology</i> , 2007, 73, 1376-1383.	4.4	48
23	COG-7-deficient Human Fibroblasts Exhibit Altered Recycling of Golgi Proteins. <i>Molecular Biology of the Cell</i> , 2006, 17, 2312-2321.	2.1	61
24	The iminosugar isofagomine increases the activity of N370S mutant acid beta-glucosidase in Gaucher fibroblasts by several mechanisms. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2006, 103, 13813-13818.	7.1	198
25	A splicing mutation in the $\beta$ -GlcNAc-1-phosphotransferase gene results in an adult onset form of mucopolipidosis III associated with sensory neuropathy and cardiomyopathy. <i>American Journal of Medical Genetics, Part A</i> , 2005, 132A, 369-375.	1.2	36
26	Identification of the Minimal Lysosomal Enzyme Recognition Domain in Cathepsin D. <i>Journal of Biological Chemistry</i> , 2005, 280, 33318-33323.	3.4	34