

# Konrad Sandhoff

## List of Publications by Year in descending order

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285  
papers

23,048  
citations

6233

80  
h-index

10424

139  
g-index

303  
all docs

303  
docs citations

303  
times ranked

16944  
citing authors

#	ARTICLE	IF	CITATIONS
1	A case of spastic paraplegia type 11 mimicking a GM2-gangliosidosis. <i>Neurological Sciences</i> , 2022, 43, 2849-2852.	0.9	0
2	Acid Sphingomyelinase, a Lysosomal and Secretory Phospholipase C, Is Key for Cellular Phospholipid Catabolism. <i>International Journal of Molecular Sciences</i> , 2021, 22, 9001.	1.8	21
3	Secondary ganglioside GM2 accumulation in mucopolysaccharidoses. <i>Molecular Genetics and Metabolism Reports</i> , 2021, 28, 100789.	0.4	0
4	Mechanism of Secondary Ganglioside and Lipid Accumulation in Lysosomal Disease. <i>International Journal of Molecular Sciences</i> , 2020, 21, 2566.	1.8	52
5	Acid ceramidase deficiency: Farber lipogranulomatosis, spinal muscular atrophy associated with progressive myoclonic epilepsy and peripheral osteolysis. , 2020, , 547-557.		0
6	Emerging mechanisms of drug-induced phospholipidosis. <i>Biological Chemistry</i> , 2019, 401, 31-46.	1.2	97
7	Lysosomal Glycosphingolipid Storage Diseases. <i>Annual Review of Biochemistry</i> , 2019, 88, 461-485.	5.0	78
8	Ganglioside GM2 catabolism is inhibited by storage compounds of mucopolysaccharidoses and by cationic amphiphilic drugs. <i>Molecular Genetics and Metabolism</i> , 2019, 128, 75-83.	0.5	12
9	Membrane lipids and their degradation compounds control GM2 catabolism at intralysosomal luminal vesicles. <i>Journal of Lipid Research</i> , 2019, 60, 1099-1111.	2.0	15
10	In Human and Mouse Spino-Cerebellar Tissue, Ataxin-2 Expansion Affects Ceramide-Sphingomyelin Metabolism. <i>International Journal of Molecular Sciences</i> , 2019, 20, 5854.	1.8	19
11	Ceramide Synthase Schlank Is a Transcriptional Regulator Adapting Gene Expression to Energy Requirements. <i>Cell Reports</i> , 2018, 22, 967-978.	2.9	40
12	Inactivation of ceramide synthase 2 catalytic activity in mice affects transcription of genes involved in lipid metabolism and cell division. <i>Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids</i> , 2018, 1863, 734-749.	1.2	16
13	Unbalanced lipolysis results in lipotoxicity and mitochondrial damage in peroxisome-deficient <i>Pex19</i> mutants. <i>Molecular Biology of the Cell</i> , 2018, 29, 396-407.	0.9	40
14	Emerging concepts of ganglioside metabolism. <i>FEBS Letters</i> , 2018, 592, 3835-3864.	1.3	72
15	Ganglioside Metabolism in Health and Disease. <i>Progress in Molecular Biology and Translational Science</i> , 2018, 156, 1-62.	0.9	55
16	Ganglioside Metabolism and Its Inherited Diseases. <i>Methods in Molecular Biology</i> , 2018, 1804, 97-141.	0.4	20
17	Lipids regulate the hydrolysis of membrane bound glucosylceramide by lysosomal $\beta$ -glucocerebrosidase. <i>Journal of Lipid Research</i> , 2017, 58, 563-577.	2.0	38
18	Synthetic Glycoforms Reveal Carbohydrate-Dependent Bioactivity of Human Saposinâ€¦D. <i>Angewandte Chemie</i> , 2017, 129, 5336-5341.	1.6	13

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19	Synthetic Glycoforms Reveal Carbohydrate-Dependent Bioactivity of Human Saposin...D. <i>Angewandte Chemie - International Edition</i> , 2017, 56, 5252-5257.	7.2	33
20	Characterization of <i>Drosophila</i> saposin-related mutants as a model for lysosomal sphingolipid storage diseases. <i>DMM Disease Models and Mechanisms</i> , 2017, 10, 737-750.	1.2	13
21	Identification of a feedback loop involving $\beta$ -glucosidase 2 and its product sphingosine sheds light on the molecular mechanisms in Gaucher disease. <i>Journal of Biological Chemistry</i> , 2017, 292, 6177-6189.	1.6	22
22	Neuronal sphingolipidoses: Membrane lipids and sphingolipid activator proteins regulate lysosomal sphingolipid catabolism. <i>Biochimie</i> , 2016, 130, 146-151.	1.3	35
23	Acid Ceramidase Deficiency. , 2015, , 395-402.		2
24	Accumulation of Glucosylceramide in the Absence of the Beta-Glucosidase GBA2 Alters Cytoskeletal Dynamics. <i>PLoS Genetics</i> , 2015, 11, e1005063.	1.5	39
25	Membrane-spanning lipids for an uncompromised monitoring of membrane fusion and intermembrane lipid transfer. <i>Journal of Lipid Research</i> , 2015, 56, 1861-1879.	2.0	19
26	Membrane lipids regulate ganglioside GM2 catabolism and GM2 activator protein activity. <i>Journal of Lipid Research</i> , 2015, 56, 1747-1761.	2.0	32
27	Acid sphingomyelinase activity is regulated by membrane lipids and facilitates cholesterol transfer by NPC2. <i>Journal of Lipid Research</i> , 2014, 55, 2606-2619.	2.0	65
28	Activation of Nrf2 in keratinocytes causes chloracne (MADISH)-like skin disease in mice. <i>EMBO Molecular Medicine</i> , 2014, 6, 442-457.	3.3	81
29	Labeled chemical biology tools for investigating sphingolipid metabolism, trafficking and interaction with lipids and proteins. <i>Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids</i> , 2014, 1841, 1161-1173.	1.2	24
30	Sphingolipids and lysosomal pathologies. <i>Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids</i> , 2014, 1841, 799-810.	1.2	88
31	The role of sphingolipid metabolism in cutaneous permeability barrier formation. <i>Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids</i> , 2014, 1841, 441-452.	1.2	141
32	Partial synthesis of ganglioside and lysoganglioside lipofoms as internal standards for MS quantification. <i>Journal of Lipid Research</i> , 2014, 55, 2692-2706.	2.0	10
33	CLN3 Loss Disturbs Membrane Microdomain Properties and Protein Transport in Brain Endothelial Cells. <i>Journal of Neuroscience</i> , 2013, 33, 18065-18079.	1.7	53
34	Functional and genetic characterization of the non-lysosomal glucosylceramidase 2 as a modifier for Gaucher disease. <i>Orphanet Journal of Rare Diseases</i> , 2013, 8, 151.	1.2	24
35	Killing from the inside. <i>Nature</i> , 2013, 502, 312-313.	13.7	79
36	Loss of CB1 receptors leads to decreased cathepsin D levels and accelerated lipofuscin accumulation in the hippocampus. <i>Mechanisms of Ageing and Development</i> , 2013, 134, 391-399.	2.2	27

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37	Gangliosides and Gangliosidoses: Principles of Molecular and Metabolic Pathogenesis. <i>Journal of Neuroscience</i> , 2013, 33, 10195-10208.	1.7	356
38	Metabolic and cellular bases of sphingolipidoses. <i>Biochemical Society Transactions</i> , 2013, 41, 1562-1568.	1.6	47
39	TCF/Lef1-Mediated Control of Lipid Metabolism Regulates Skin Barrier Function. <i>Journal of Investigative Dermatology</i> , 2012, 132, 337-345.	0.3	21
40	Ablation of Neuronal Ceramide Synthase 1 in Mice Decreases Ganglioside Levels and Expression of Myelin-associated Glycoprotein in Oligodendrocytes. <i>Journal of Biological Chemistry</i> , 2012, 287, 41888-41902.	1.6	117
41	My journey into the world of sphingolipids and sphingolipidoses. <i>Proceedings of the Japan Academy Series B: Physical and Biological Sciences</i> , 2012, 88, 554-582.	1.6	35
42	Nrf2 links epidermal barrier function with antioxidant defense. <i>EMBO Molecular Medicine</i> , 2012, 4, 364-379.	3.3	153
43	Mass spectrometric analysis of neutral sphingolipids: Methods, applications, and limitations. <i>Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids</i> , 2011, 1811, 854-860.	1.2	35
44	Regulation of the NPC2 protein-mediated cholesterol trafficking by membrane lipids. <i>Journal of Neurochemistry</i> , 2011, 116, 702-707.	2.1	43
45	Role for LAMP-2 in endosomal cholesterol transport. <i>Journal of Cellular and Molecular Medicine</i> , 2011, 15, 280-295.	1.6	70
46	Biological Function of the Cellular Lipid BMP as a Key Activator for Cholesterol Sorting and Membrane Digestion. <i>Neurochemical Research</i> , 2011, 36, 1594-1600.	1.6	108
47	PAR2 absence completely rescues inflammation and ichthyosis caused by altered CAP1/Prss8 expression in mouse skin. <i>Nature Communications</i> , 2011, 2, 161.	5.8	96
48	Lysosomal Lipid Storage Diseases. <i>Cold Spring Harbor Perspectives in Biology</i> , 2011, 3, a004804-a004804.	2.3	142
49	Sphingolipid Storage Affects Autophagic Metabolism of the Amyloid Precursor Protein and Promotes A $\beta$ Generation. <i>Journal of Neuroscience</i> , 2011, 31, 1837-1849.	1.7	82
50	Lysosomal degradation of membrane lipids. <i>FEBS Letters</i> , 2010, 584, 1700-1712.	1.3	229
51	Hsp70 stabilizes lysosomes and reverts Niemann-Pick disease-associated lysosomal pathology. <i>Nature</i> , 2010, 463, 549-553.	13.7	425
52	Golgi-to-phagosome transport of acid sphingomyelinase and prosaposin is mediated by sortilin. <i>Journal of Cell Science</i> , 2010, 123, 2502-2511.	1.2	70
53	Role of endosomal membrane lipids and NPC2 in cholesterol transfer and membrane fusion. <i>Journal of Lipid Research</i> , 2010, 51, 1747-1760.	2.0	71
54	Prion-induced Activation of Cholesterogenic Gene Expression by Srebp2 in Neuronal Cells. <i>Journal of Biological Chemistry</i> , 2009, 284, 31260-31269.	1.6	34

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55	Adult Ceramide Synthase 2 (CERS2)-deficient Mice Exhibit Myelin Sheath Defects, Cerebellar Degeneration, and Hepatocarcinomas. <i>Journal of Biological Chemistry</i> , 2009, 284, 33549-33560.	1.6	245
56	Saposin B-dependent Reconstitution of Arylsulfatase A Activity in Vitro and in Cell Culture Models of Metachromatic Leukodystrophy. <i>Journal of Biological Chemistry</i> , 2009, 284, 9372-9381.	1.6	17
57	Membrane lipids and degenerative lysosomal storage disorders. <i>Clinical Therapeutics</i> , 2009, 31, S179-S180.	1.1	1
58	Principles of lysosomal membrane degradation. <i>Biochimica Et Biophysica Acta - Molecular Cell Research</i> , 2009, 1793, 674-683.	1.9	203
59	Normal phase liquid chromatography coupled to quadrupole time of flight atmospheric pressure chemical ionization mass spectrometry for separation, detection and mass spectrometric profiling of neutral sphingolipids and cholesterol. <i>Journal of Chromatography B: Analytical Technologies in the Biomedical and Life Sciences</i> , 2009, 877, 2976-2982.	1.2	54
60	Hexosaminidase assays. <i>Glycoconjugate Journal</i> , 2009, 26, 945-952.	1.4	66
61	Schlank, a member of the ceramide synthase family controls growth and body fat in <i>Drosophila</i> . <i>EMBO Journal</i> , 2009, 28, 3706-3716.	3.5	76
62	Direct observation of the nanoscale dynamics of membrane lipids in a living cell. <i>Nature</i> , 2009, 457, 1159-1162.	13.7	1,392
63	Neural precursor cell cultures from GM2 gangliosidosis animal models recapitulate the biochemical and molecular hallmarks of the brain pathology. <i>Journal of Neurochemistry</i> , 2009, 109, 135-147.	2.1	38
64	Crystal Structures of Human Saposins C and D: Implications for Lipid Recognition and Membrane Interactions. <i>Structure</i> , 2008, 16, 809-817.	1.6	78
65	Insulin receptor and lipid metabolism pathology in ataxin-2 knock-out mice. <i>Human Molecular Genetics</i> , 2008, 17, 1465-1481.	1.4	107
66	Postnatal Requirement of the Epithelial Sodium Channel for Maintenance of Epidermal Barrier Function. <i>Journal of Biological Chemistry</i> , 2008, 283, 2622-2630.	1.6	38
67	Integrity and Barrier Function of the Epidermis Critically Depend on Glucosylceramide Synthesis. <i>Journal of Biological Chemistry</i> , 2007, 282, 3083-3094.	1.6	105
68	Development of an assay for the intermembrane transfer of cholesterol by Niemann-Pick C2 protein. <i>Biological Chemistry</i> , 2007, 388, 617-26.	1.2	60
69	Overexpression and mass spectrometry analysis of mature human acid ceramidase. <i>Biological Chemistry</i> , 2007, 388, 1333-1343.	1.2	29
70	Identification of the protein receptor binding site of botulinum neurotoxins B and G proves the double-receptor concept. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2007, 104, 359-364.	3.3	169
71	Separation and mass spectrometric characterization of covalently bound skin ceramides using LC/APCI-MS and Nano-ESI-MS/MS. <i>Journal of Chromatography B: Analytical Technologies in the Biomedical and Life Sciences</i> , 2007, 852, 562-570.	1.2	56
72	Normal epidermal differentiation but impaired skin-barrier formation upon keratinocyte-restricted IKK1 ablation. <i>Nature Cell Biology</i> , 2007, 9, 461-469.	4.6	87

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73	Saposin $\alpha$ mobilizes lipids from cholesterol-poor and bis(monoacylglycero)phosphate-rich membranes at acidic pH. <i>FEBS Journal</i> , 2007, 274, 3405-3420.	2.2	58
74	Optimization of submerged keratinocyte cultures for the synthesis of barrier ceramides. <i>European Journal of Cell Biology</i> , 2007, 86, 657-673.	1.6	25
75	Characterization of Human Saposins by NMR Spectroscopy. <i>Biochemistry</i> , 2006, 45, 5206-5216.	1.2	27
76	Sphingolipid metabolism diseases. <i>Biochimica Et Biophysica Acta - Biomembranes</i> , 2006, 1758, 2057-2079.	1.4	306
77	Apoptotic Vesicles Crossprime CD8 T Cells and Protect against Tuberculosis. <i>Immunity</i> , 2006, 24, 105-117.	6.6	353
78	Crystallization and preliminary characterization of three different crystal forms of human saposin C heterologously expressed in <i>Pichia pastoris</i> . <i>Acta Crystallographica Section F: Structural Biology Communications</i> , 2006, 62, 117-120.	0.7	5
79	The enzyme-binding region of human GM2-activator protein. <i>FEBS Journal</i> , 2006, 273, 982-991.	2.2	28
80	Saposin A Mobilizes Lipids from Low Cholesterol and High Bis(monoacylglycerol)phosphate-containing Membranes. <i>Journal of Biological Chemistry</i> , 2006, 281, 32451-32460.	1.6	69
81	Saposin C stimulates growth and invasion, activates p42/44 and SAPK/JNK signaling pathways of MAPK and upregulates uPA/uPAR expression in prostate cancer and stromal cells. <i>Asian Journal of Andrology</i> , 2005, 7, 147-158.	0.8	26
82	Amplification and overexpression of prosaposin in prostate cancer. <i>Genes Chromosomes and Cancer</i> , 2005, 44, 351-364.	1.5	46
83	Site-Specific Cleavage-A Model System for the Identification of Lipid-Modified Glutamate Residues in Proteins. <i>ChemBioChem</i> , 2005, 6, 178-185.	1.3	5
84	Synthesis of novel NBD-GM1 and NBD-GM2 for the transfer activity of GM2-activator protein by a FRET-based assay system. <i>Glycobiology</i> , 2005, 15, 1302-1311.	1.3	51
85	Interruption of ganglioside synthesis produces central nervous system degeneration and altered axon-glia interactions. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2005, 102, 2725-2730.	3.3	212
86	Inhibition of Glycosphingolipid Biosynthesis Reduces Secretion of the $\beta$ -Amyloid Precursor Protein and Amyloid $\beta$ -Peptide. <i>Journal of Biological Chemistry</i> , 2005, 280, 28110-28117.	1.6	115
87	The epidermal barrier function is dependent on the serine protease CAP1/Prss8. <i>Journal of Cell Biology</i> , 2005, 170, 487-496.	2.3	255
88	Acid Ceramidase Overexpression Prevents the Inhibitory Effects of Saturated Fatty Acids on Insulin Signaling. <i>Journal of Biological Chemistry</i> , 2005, 280, 20148-20153.	1.6	188
89	A novel mass spectrometric assay for the cerebroside sulfate activator protein (saposin B) and arylsulfatase A. <i>Journal of Lipid Research</i> , 2005, 46, 2254-2264.	2.0	9
90	Lipid-binding Proteins in Membrane Digestion, Antigen Presentation, and Antimicrobial Defense. <i>Journal of Biological Chemistry</i> , 2005, 280, 41125-41128.	1.6	70

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91	PRINCIPLES OF LYSOSOMAL MEMBRANE DIGESTION: Stimulation of Sphingolipid Degradation by Sphingolipid Activator Proteins and Anionic Lysosomal Lipids. <i>Annual Review of Cell and Developmental Biology</i> , 2005, 21, 81-103.	4.0	397
92	Functional characterization of the postulated intramolecular sphingolipid activator protein domain of human acid sphingomyelinase. <i>Biological Chemistry</i> , 2004, 385, 1193-5.	1.2	21
93	Photoaffinity labelling of the Human GM2-activator protein. Mechanistic insight into ganglioside GM2 degradation. <i>FEBS Journal</i> , 2004, 271, 614-627.	0.2	49
94	Saposin C is required for lipid presentation by human CD1b. <i>Nature Immunology</i> , 2004, 5, 169-174.	7.0	160
95	Nitric oxide regulates synthesis of gene products involved in keratinocyte differentiation and ceramide metabolism. <i>European Journal of Cell Biology</i> , 2004, 83, 667-679.	1.6	16
96	Loss of keratin 10 is accompanied by increased sebocyte proliferation and differentiation. <i>European Journal of Cell Biology</i> , 2004, 83, 747-759.	1.6	38
97	Prosaptide TX14A stimulates growth, migration, and invasion and activates the Raf-MEK-ERK-RSK-Elk-1 signaling pathway in prostate cancer cells. <i>Prostate</i> , 2004, 61, 114-123.	1.2	23
98	Expression of the GM2-activator protein in the methylotrophic yeast <i>Pichia pastoris</i> , purification, isotopic labeling, and biophysical characterization. <i>Protein Expression and Purification</i> , 2004, 34, 147-157.	0.6	23
99	Interactions of acid sphingomyelinase and lipid bilayers in the presence of the tricyclic antidepressant desipramine. <i>FEBS Letters</i> , 2004, 559, 96-98.	1.3	172
100	Purified recombinant human prosaposin forms oligomers that bind procathepsin D and affect its autoactivation. <i>Biochemical Journal</i> , 2004, 383, 507-515.	1.7	63
101	Characterization of two Turkish $\beta$ -hexosaminidase mutations causing Tay-Sachs disease. <i>Brain and Development</i> , 2003, 25, 191-194.	0.6	13
102	A new point mutation (G412 to A) at the last nucleotide of exon 3 of hexosaminidase $\beta$ -subunit gene affects splicing. <i>Brain and Development</i> , 2003, 25, 203-206.	0.6	11
103	Human acid sphingomyelinase. Assignment of the disulfide bond pattern. <i>FEBS Journal</i> , 2003, 270, 1076-1088.	0.2	40
104	6(S)-Methyl-3(S)-(1-methylethyl)piperazin-2-one. <i>Acta Crystallographica Section E: Structure Reports Online</i> , 2003, 59, o171-o173.	0.2	0
105	Biosynthesis and degradation of mammalian glycosphingolipids. <i>Philosophical Transactions of the Royal Society B: Biological Sciences</i> , 2003, 358, 847-861.	1.8	157
106	The X-ray Crystal Structure of Human $\beta$ -Hexosaminidase B Provides New Insights into Sandhoff Disease. <i>Journal of Molecular Biology</i> , 2003, 328, 669-681.	2.0	109
107	Physiological relevance of sphingolipid activator proteins in cultured human fibroblasts. <i>Biochimie</i> , 2003, 85, 439-448.	1.3	10
108	Detection and identification of Vav1 protein in primary cultured murine cerebellar neurons and in neuroblastoma cells (SH-SY5Y and Neuro-2a). <i>Neuroscience Letters</i> , 2003, 339, 37-40.	1.0	9

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109	Expression of recombinant human GM2-activator protein in insect cells: purification and characterization by mass spectrometry. <i>Protein Expression and Purification</i> , 2003, 27, 259-266.	0.6	27
110	The Reverse Activity of Human Acid Ceramidase. <i>Journal of Biological Chemistry</i> , 2003, 278, 29948-29953.	1.6	133
111	Enhanced insulin sensitivity in mice lacking ganglioside GM3. <i>Proceedings of the National Academy of Sciences of the United States of America</i> , 2003, 100, 3445-3449.	3.3	487
112	Recombinant Ganglioside GM2 Synthase Expression in Insect Cells and Enzyme Assay. <i>Methods in Enzymology</i> , 2003, 363, 476-489.	0.4	0
113	Purification and Characterization of Recombinant, Human Acid Ceramidase. <i>Journal of Biological Chemistry</i> , 2003, 278, 32978-32986.	1.6	88
114	Phosphatidylinositol-3,5-Bisphosphate Is a Potent and Selective Inhibitor of Acid Sphingomyelinase. <i>Biological Chemistry</i> , 2003, 384, 1293-8.	1.2	59
115	RNA Interference: Analyzing the Function of Glycoproteins and Glycosylating Proteins in Mammalian Cells. <i>Methods in Enzymology</i> , 2003, 363, 173-190.	0.4	1
116	Interaction of the GM2 Activator Protein with Sulfated and Sialylated Glycosphingolipids. <i>Methods in Enzymology</i> , 2003, 363, 207-222.	0.4	3
117	Chapter 14 Sphingolipids: metabolism and cell signaling. <i>New Comprehensive Biochemistry</i> , 2002, 36, 373-407.	0.1	48
118	Combinatorial Ganglioside Biosynthesis. <i>Journal of Biological Chemistry</i> , 2002, 277, 25859-25862.	1.6	271
119	Sphingolipid metabolism during epidermal barrier development in mice. <i>Journal of Lipid Research</i> , 2002, 43, 1727-1733.	2.0	37
120	Physiological Substrates for Human Lysosomal $\beta$ -Hexosaminidase S. <i>Journal of Biological Chemistry</i> , 2002, 277, 2562-2572.	1.6	81
121	Procathepsin D Interacts with Prosaposin in Cancer Cells but Its Internalization Is Not Mediated by LDL Receptor-Related Protein. <i>Experimental Cell Research</i> , 2002, 277, 210-219.	1.2	25
122	Insertional Mutagenesis of the Mouse Acid Ceramidase Gene Leads to Early Embryonic Lethality in Homozygotes and Progressive Lipid Storage Disease in Heterozygotes. <i>Genomics</i> , 2002, 79, 218-224.	1.3	104
123	An Inducible Mouse Model of Late Onset Tay-Sachs Disease. <i>Neurobiology of Disease</i> , 2002, 10, 201-210.	2.1	31
124	Deficiency of Epidermal Protein-Bound $\beta$ -Hydroxyceramides in Atopic Dermatitis. <i>Journal of Investigative Dermatology</i> , 2002, 119, 166-173.	0.3	212
125	Identification of ceramide binding proteins in neuronal cells: a critical point of view. <i>Neurochemical Research</i> , 2002, 27, 717-727.	1.6	22
126	Systemic inflammation in glucocerebrosidase-deficient mice with minimal glucosylceramide storage. <i>Journal of Clinical Investigation</i> , 2002, 109, 1215-1221.	3.9	114



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127	Systemic inflammation in glucocerebrosidase-deficient mice with minimal glucosylceramide storage. <i>Journal of Clinical Investigation</i> , 2002, 109, 1215-1221.	3.9	73
128	8. The GM2-gangliosidoses and the elucidation of the $\beta$ 2-hexosaminidase system. <i>Advances in Genetics</i> , 2001, 44, 67-91.	0.8	15
129	Molecular analysis of acid ceramidase deficiency in patients with Farber disease. <i>Human Mutation</i> , 2001, 17, 199-209.	1.1	76
130	Interaction of the GM2-activator protein with phospholipid-ganglioside bilayer membranes and with monolayers at the air-water interface. <i>FEBS Journal</i> , 2001, 261, 650-658.	0.2	72
131	Up-regulation of prosaposin by the retinoid HPR and the effect on ceramide production and integrin receptors. <i>FASEB Journal</i> , 2001, 15, 1475-1477.	0.2	15
132	Stimulation of Acid Sphingomyelinase Activity by Lysosomal Lipids and Sphingolipid Activator Proteins. <i>Biological Chemistry</i> , 2001, 382, 283-90.	1.2	94
133	Human Acid Ceramidase. <i>Journal of Biological Chemistry</i> , 2001, 276, 35352-35360.	1.6	98
134	Interfacial Regulation of Acid Ceramidase Activity. <i>Journal of Biological Chemistry</i> , 2001, 276, 5760-5768.	1.6	113
135	Degradation of Membrane-bound Ganglioside GM2 by $\beta$ 2-Hexosaminidase A. <i>Journal of Biological Chemistry</i> , 2001, 276, 12685-12690.	1.6	57
136	Mice Expressing Only Monosialoganglioside GM3 Exhibit Lethal Audiogenic Seizures. <i>Journal of Biological Chemistry</i> , 2001, 276, 6885-6888.	1.6	218
137	Complete analysis of the glycosylation and disulfide bond pattern of human $\beta$ -hexosaminidase B by MALDI-MS. <i>Glycobiology</i> , 2001, 11, 549-556.	1.3	30
138	CD95 Signaling via Ceramide-rich Membrane Rafts. <i>Journal of Biological Chemistry</i> , 2001, 276, 20589-20596.	1.6	559
139	Biosynthesis and Degradation. , 2001, , 2149-2181.		0
140	[10] In Vitro assays for enzymes of ganglioside synthesis. <i>Methods in Enzymology</i> , 2000, 311, 82-94.	0.4	8
141	[29] Sphingolipid hydrolases and activator proteins. <i>Methods in Enzymology</i> , 2000, 311, 255-276.	0.4	13
142	[47] Sphingolipid photoaffinity labels. <i>Methods in Enzymology</i> , 2000, 311, 568-600.	0.4	16
143	Biomolecule Function: No Reliable Prediction from Cell Culture. <i>Traffic</i> , 2000, 1, 803-804.	1.3	21
144	Characterization of sphingomyelinase activity released by thrombin-stimulated platelets. <i>Molecular and Cellular Biochemistry</i> , 2000, 205, 75-81.	1.4	38

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145	Degradation of Membrane-bound Ganglioside GM1. <i>Journal of Biological Chemistry</i> , 2000, 275, 35814-35819.	1.6	79
146	Overexpression of Acid Ceramidase Protects from Tumor Necrosis Factor-Induced Cell Death. <i>Journal of Experimental Medicine</i> , 2000, 192, 601-612.	4.2	164
147	Purification and Characterization of a Magnesium-dependent Neutral Sphingomyelinase from Bovine Brain. <i>Journal of Biological Chemistry</i> , 2000, 275, 7641-7647.	1.6	71
148	Characterization of Regulatory Elements in the 5'-Flanking Region of the GM2 Activator Gene. <i>Biological Chemistry</i> , 2000, 381, 531-44.	1.2	5
149	Pathology of Glycosphingolipid Metabolism: The Molecular and Cellular Basis of Neurodegenerative Disease. <i>Neuroscientist</i> , 2000, 6, 285-296.	2.6	1
150	Characterization of two $\beta$ -galactosidase mutants (Q279E and R301Q) found in an atypical variant of Fabry disease. <i>Biochimica Et Biophysica Acta - Molecular Basis of Disease</i> , 2000, 1501, 227-235.	1.8	36
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