## **Konrad Sandhoff**

List of Publications by Year in descending order

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285 papers 23,048 citations

80 h-index 139 g-index

303 all docs 303 docs citations

times ranked

303

16944 citing authors

#	Article	IF	Citations
1	Direct observation of the nanoscale dynamics of membrane lipids in a living cell. Nature, 2009, 457, 1159-1162.	13.7	1,392
2	CD95 Signaling via Ceramide-rich Membrane Rafts. Journal of Biological Chemistry, 2001, 276, 20589-20596.	1.6	559
3	Enhanced insulin sensitivity in mice lacking ganglioside GM3. Proceedings of the National Academy of Sciences of the United States of America, 2003, 100, 3445-3449.	3.3	487
4	Acid sphingomyelinase deficient mice: a model of types A and B Niemann–Pick disease. Nature Genetics, 1995, 10, 288-293.	9.4	457
5	Hsp70 stabilizes lysosomes and reverts Niemann–Pick disease-associated lysosomal pathology. Nature, 2010, 463, 549-553.	13.7	425
6	Mouse models of Tay–Sachs and Sandhoff diseases differ in neurologic phenotype and ganglioside metabolism. Nature Genetics, 1995, 11, 170-176.	9.4	411
7	PRINCIPLES OF LYSOSOMAL MEMBRANE DIGESTION: Stimulation of Sphingolipid Degradation by Sphingolipid Activator Proteins and Anionic Lysosomal Lipids. Annual Review of Cell and Developmental Biology, 2005, 21, 81-103.	4.0	397
8	Physiology and pathophysiology of sphingolipid metabolism and signaling. Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids, 2000, 1485, 63-99.	1.2	372
9	Sphingolipids—Their Metabolic Pathways and the Pathobiochemistry of Neurodegenerative Diseases. Angewandte Chemie - International Edition, 1999, 38, 1532-1568.	7.2	360
10	Gangliosides and Gangliosidoses: Principles of Molecular and Metabolic Pathogenesis. Journal of Neuroscience, 2013, 33, 10195-10208.	1.7	356
11	Apoptotic Vesicles Crossprime CD8 T Cells and Protect against Tuberculosis. Immunity, 2006, 24, 105-117.	6.6	353
12	Acidic Sphingomyelinase Mediates Entry of N. gonorrhoeae into Nonphagocytic Cells. Cell, 1997, 91, 605-615.	13.5	307
13	Sphingolipid metabolism diseases. Biochimica Et Biophysica Acta - Biomembranes, 2006, 1758, 2057-2079.	1.4	306
14	Combinatorial Ganglioside Biosynthesis. Journal of Biological Chemistry, 2002, 277, 25859-25862.	1.6	271
15	Characterization of Ceramide Synthesis. Journal of Biological Chemistry, 1997, 272, 22432-22437.	1.6	266
16	The epidermal barrier function is dependent on the serine protease CAP1/Prss8. Journal of Cell Biology, 2005, 170, 487-496.	2.3	255
17	Activator proteins and topology of lysosomal sphingolipid catabolism. Lipids and Lipid Metabolism, 1992, 1126, 1-16.	2.6	254
18	Adult Ceramide Synthase 2 (CERS2)-deficient Mice Exhibit Myelin Sheath Defects, Cerebellar Degeneration, and Hepatocarcinomas. Journal of Biological Chemistry, 2009, 284, 33549-33560.	1.6	245

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19	Molecular Cloning and Characterization of a Full-length Complementary DNA Encoding Human Acid Ceramidase. Journal of Biological Chemistry, 1996, 271, 33110-33115.	1.6	232
20	Lysosomal degradation of membrane lipids. FEBS Letters, 2010, 584, 1700-1712.	1.3	229
21	The Tricyclic Antidepressant Desipramine Causes Proteolytic Degradation of Lysosomal Sphingomyelinase in Human Fibroblasts. Biological Chemistry Hoppe-Seyler, 1994, 375, 447-450.	1.4	220
22	Mice Expressing Only Monosialoganglioside GM3 Exhibit Lethal Audiogenic Seizures. Journal of Biological Chemistry, 2001, 276, 6885-6888.	1.6	218
23	Deficiency of Epidermal Protein-Bound ω-Hydroxyceramides in Atopic Dermatitis. Journal of Investigative Dermatology, 2002, 119, 166-173.	0.3	212
24	Interruption of ganglioside synthesis produces central nervous system degeneration and altered axon-glial interactions. Proceedings of the National Academy of Sciences of the United States of America, 2005, 102, 2725-2730.	3.3	212
25	Principles of lysosomal membrane degradation. Biochimica Et Biophysica Acta - Molecular Cell Research, 2009, 1793, 674-683.	1.9	203
26	Mice lacking both subunits of lysosomal $\hat{l}^2\hat{a}\in$ hexosaminidase display gangliosidosis and mucopolysaccharidosis. Nature Genetics, 1996, 14, 348-352.	9.4	194
27	Purification, Characterization, and Biosynthesis of Human Acid Ceramidase. Journal of Biological Chemistry, 1995, 270, 11098-11102.	1.6	193
28	Acid Ceramidase Overexpression Prevents the Inhibitory Effects of Saturated Fatty Acids on Insulin Signaling. Journal of Biological Chemistry, 2005, 280, 20148-20153.	1.6	188
29	Metabolism and intracellular transport of glycosphingolipids. Biochemistry, 1990, 29, 10865-10871.	1.2	178
30	Purification and Characterization of an Activator Protein for the Degradation of Glycolipids GM2and GA2by Hexosaminidase A. Hoppe-Seyler's Zeitschrift Fýr Physiologische Chemie, 1979, 360, 1837-1850.	1.7	176
31	Interactions of acid sphingomyelinase and lipid bilayers in the presence of the tricyclic antidepressant desipramine. FEBS Letters, 2004, 559, 96-98.	1.3	172
32	Identification of the protein receptor binding site of botulinum neurotoxins B and G proves the double-receptor concept. Proceedings of the National Academy of Sciences of the United States of America, 2007, 104, 359-364.	3.3	169
33	Overexpression of Acid Ceramidase Protects from Tumor Necrosis Factor–Induced Cell Death. Journal of Experimental Medicine, 2000, 192, 601-612.	4.2	164
34	Topology of glycosphingolipid degradation. Trends in Cell Biology, 1996, 6, 98-103.	3.6	161
35	Incorporation of ganglioside analogs into fibroblast cell membranes. A spin-label study. Biochemistry, 1983, 22, 5041-5048.	1.2	160
36	Saposin C is required for lipid presentation by human CD1b. Nature Immunology, 2004, 5, 169-174.	7.0	160

3

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37	Biosynthesis and degradation of mammalian glycosphingolipids. Philosophical Transactions of the Royal Society B: Biological Sciences, 2003, 358, 847-861.	1.8	157
38	Incorporation and metabolism of ganglioside GM2 in skin fibroblasts from normal and GM2 gangliosidosis subjects. FEBS Journal, 1985, 149, 247-255.	0.2	155
39	Mutation in the sphingolipid activator protein 2 in a patient with a variant of Gaucher disease. FEBS Letters, 1991, 284, 57-59.	1.3	153
40	Nrf2 links epidermal barrier function with antioxidant defense. EMBO Molecular Medicine, 2012, 4, 364-379.	3.3	153
41	A genetic model of substrate deprivation therapy for a glycosphingolipid storage disorder. Journal of Clinical Investigation, 1999, 103, 497-505.	3.9	153
42	Lysosomal Enzyme Precursors in Human Fibroblasts. Activation of Cathepsin D Precursor in vitro and Activity of beta-Hexosaminidase A Precursor towards Ganglioside GM2. FEBS Journal, 1982, 125, 317-321.	0.2	152
43	Complexing of Glycolipids and Their Transfer between Membranes by the Activator Protein for Degradation of Lysosomal Ganglioside GM2. FEBS Journal, 1982, 123, 455-464.	0.2	151
44	Lysosomal Lipid Storage Diseases. Cold Spring Harbor Perspectives in Biology, 2011, 3, a004804-a004804.	2.3	142
45	The role of sphingolipid metabolism in cutaneous permeabilitybarrier formation. Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids, 2014, 1841, 441-452.	1.2	141
46	Lysosomal Degradation on Vesicular Membrane Surfaces. Journal of Biological Chemistry, 1998, 273, 30271-30278.	1.6	140
47	Biosynthesis of sphingolipids: Dihydroceramide and not sphinganine is desaturated by cultured cells. Biochemical and Biophysical Research Communications, 1992, 189, 14-20.	1.0	139
48	Sphingolipid Activator Proteins Are Required for Epidermal Permeability Barrier Formation. Journal of Biological Chemistry, 1999, 274, 11038-11045.	1.6	138
49	Structure of Full-Length cDNA Coding for Sulfatide Activator, a Co- $\hat{l}^2$ -Glucosidase and Two Other Homologous Proteins: Two Alternate Forms of the Sulfatide Activator 1. Journal of Biochemistry, 1989, 105, 152-154.	0.9	137
50	The Reverse Activity of Human Acid Ceramidase. Journal of Biological Chemistry, 2003, 278, 29948-29953.	1.6	133
51	The Human Acid Ceramidase Gene (ASAH): Structure, Chromosomal Location, Mutation Analysis, and Expression. Genomics, 1999, 62, 223-231.	1.3	130
52	Stereoselective synthesis of î±- C-allyl-glycopyranosides. Tetrahedron Letters, 1985, 26, 1479-1482.	0.7	129
53	Acid sphingomyelinase from human urine: purification and characterization. Lipids and Lipid Metabolism, 1987, 922, 323-336.	2.6	129
54	Fas/CD95/Apo-I activates the acidic sphingomyelinase via Caspases. Cell Death and Differentiation, 1998, 5, 29-37.	5.0	128

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55	LiBH4(NaBH4)/Me3SiCl, an Unusually Strong and Versatile Reducing Agent. Angewandte Chemie International Edition in English, 1989, 28, 218-220.	4.4	124
56	Modulation of Ganglioside Biosynthesis in Primary Cultured Neurons. Journal of Neurochemistry, 1989, 52, 207-214.	2.1	123
57	Ablation of Neuronal Ceramide Synthase 1 in Mice Decreases Ganglioside Levels and Expression of Myelin-associated Glycoprotein in Oligodendrocytes. Journal of Biological Chemistry, 2012, 287, 41888-41902.	1.6	117
58	Inhibition of Glycosphingolipid Biosynthesis Reduces Secretion of the $\hat{l}^2$ -Amyloid Precursor Protein and Amyloid $\hat{l}^2$ -Peptide*[boxs]. Journal of Biological Chemistry, 2005, 280, 28110-28117.	1.6	115
59	Systemic inflammation in glucocerebrosidase-deficient mice with minimal glucosylceramide storage. Journal of Clinical Investigation, 2002, 109, 1215-1221.	3.9	114
60	Interfacial Regulation of Acid Ceramidase Activity. Journal of Biological Chemistry, 2001, 276, 5760-5768.	1.6	113
61	Cloning and Characterization of the Full-Length cDNA and Genomic Sequences Encoding Murine Acid Ceramidase. Genomics, 1998, 50, 267-274.	1.3	109
62	The X-ray Crystal Structure of Human β-Hexosaminidase B Provides New Insights into Sandhoff Disease. Journal of Molecular Biology, 2003, 328, 669-681.	2.0	109
63	Biological Function of the Cellular Lipid BMP—BMP as a Key Activator for Cholesterol Sorting and Membrane Digestion. Neurochemical Research, 2011, 36, 1594-1600.	1.6	108
64	Insulin receptor and lipid metabolism pathology in ataxin-2 knock-out mice. Human Molecular Genetics, 2008, 17, 1465-1481.	1.4	107
65	Anreicherung und Charakterisierung zweier Formen der menschlichen N-Acetyl-Î <sup>2</sup> -D-hexosaminidase. Hoppe-Seyler's Zeitschrift Für Physiologische Chemie, 1971, 352, 1119-1133.	1.7	105
66	[26] Lysogangliosides: Synthesis and use in preparing labeled gangliosides. Methods in Enzymology, 1987, 138, 319-341.	0.4	105
67	Integrity and Barrier Function of the Epidermis Critically Depend on Glucosylceramide Synthesis. Journal of Biological Chemistry, 2007, 282, 3083-3094.	1.6	105
68	The Precursor of Sulfatide Activator Protein is Processed to Three Different Proteins. Biological Chemistry Hoppe-Seyler, 1988, 369, 317-328.	1.4	104
69	Insertional Mutagenesis of the Mouse Acid Ceramidase Gene Leads to Early Embryonic Lethality in Homozygotes and Progressive Lipid Storage Disease in Heterozygotes. Genomics, 2002, 79, 218-224.	1.3	104
70	Human Acid Ceramidase. Journal of Biological Chemistry, 2001, 276, 35352-35360.	1.6	98
71	Emerging mechanisms of drug-induced phospholipidosis. Biological Chemistry, 2019, 401, 31-46.	1.2	97
72	PAR2 absence completely rescues inflammation and ichthyosis caused by altered CAP1/Prss8 expression in mouse skin. Nature Communications, 2011, 2, 161.	5.8	96

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73	Stimulation of Acid Sphingomyelinase Activity by Lysosomal Lipids and Sphingolipid Activator Proteins. Biological Chemistry, 2001, 382, 283-90.	1.2	94
74	Specificity of Human Liver Hexosaminidases A and B against Glycosphingolipids G <sub>M2</sub> and G <sub>A2</sub> . Purification of the Enzymes by Affinity Chromatography Employing Specific Elution. Hoppe-Seyler's Zeitschrift Fýr Physiologische Chemie, 1977, 358, 779-788.	1.7	89
75	Purification and Characterization of Recombinant, Human Acid Ceramidase. Journal of Biological Chemistry, 2003, 278, 32978-32986.	1.6	88
76	Sphingolipids and lysosomal pathologies. Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids, 2014, 1841, 799-810.	1.2	88
77	Normal epidermal differentiation but impaired skin-barrier formation upon keratinocyte-restricted IKK1 ablation. Nature Cell Biology, 2007, 9, 461-469.	4.6	87
78	On a biochemically special form of infantile amaurotic idiocy. Biochimica Et Biophysica Acta, 1963, 70, 354-356.	1.3	86
79	Accumulation of protein-bound epidermal glucosylceramides in $\hat{l}^2$ -glucocerebrosidase deficient type 2 Gaucher mice. FEBS Letters, 1999, 447, 167-170.	1.3	86
80	Sphingolipid Storage Affects Autophagic Metabolism of the Amyloid Precursor Protein and Promotes Al <sup>2</sup> Generation. Journal of Neuroscience, 2011, 31, 1837-1849.	1.7	82
81	Physiological Substrates for Human Lysosomal Î <sup>2</sup> -Hexosaminidase S. Journal of Biological Chemistry, 2002, 277, 2562-2572.	1.6	81
82	Activation of Nrf2 in keratinocytes causes chloracne (MADISH)â€like skin disease in mice. EMBO Molecular Medicine, 2014, 6, 442-457.	3.3	81
83	Degradation of Membrane-bound Ganglioside GM1. Journal of Biological Chemistry, 2000, 275, 35814-35819.	1.6	79
84	Killing from the inside. Nature, 2013, 502, 312-313.	13.7	79
85	Crystal Structures of Human Saposins C and D: Implications for Lipid Recognition and Membrane Interactions. Structure, 2008, 16, 809-817.	1.6	78
86	Lysosomal Glycosphingolipid Storage Diseases. Annual Review of Biochemistry, 2019, 88, 461-485.	5.0	78
87	Molecular analysis of acid ceramidase deficiency in patients with Farber disease. Human Mutation, 2001, 17, 199-209.	1.1	76
88	Schlank, a member of the ceramide synthase family controls growth and body fat in Drosophila. EMBO Journal, 2009, 28, 3706-3716.	3.5	76
89	Direct evidence by carbon-13 NMR spectroscopy for the erythro configuration of the sphingoid moiety in Gaucher cerebroside and other natural sphingolipids. FEBS Journal, 1985, 146, 59-64.	0.2	73
90	Recent Advances in the Biochemistry of Sphingolipidoses. Brain Pathology, 1998, 8, 79-100.	2.1	73

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91	Systemic inflammation in glucocerebrosidase-deficient mice with minimal glucosylceramide storage. Journal of Clinical Investigation, 2002, 109, 1215-1221.	3.9	73
92	Genomic Organization of the Gene Coding for TIRC7, a Novel Membrane Protein Essential for T Cell Activation. Genomics, 1999, 57, 398-406.	1.3	72
93	Interaction of the GM2-activator protein with phospholipid-ganglioside bilayer membranes and with monolayers at the air-water interface. FEBS Journal, 2001, 261, 650-658.	0.2	72
94	Emerging concepts of ganglioside metabolism. FEBS Letters, 2018, 592, 3835-3864.	1.3	72
95	Pyrene-labeled gangliosides: micelle formation in aqueous solution, lateral diffusion, and thermotropic behavior in phosphatidylcholine bilayers. Biochemistry, 1987, 26, 5943-5952.	1.2	71
96	cis-4-Methylsphingosine Decreases Sphingolipid Biosynthesis by Specifically Interfering with Serine Palmitoyltransferase Activity in Primary Cultured Neurons. Journal of Biological Chemistry, 1997, 272, 15825-15833.	1.6	71
97	Purification and Characterization of a Magnesium-dependent Neutral Sphingomyelinase from Bovine Brain. Journal of Biological Chemistry, 2000, 275, 7641-7647.	1.6	71
98	Role of endosomal membrane lipids and NPC2 in cholesterol transfer and membrane fusion. Journal of Lipid Research, 2010, 51, 1747-1760.	2.0	71
99	A mutation in the gene of a glycolipid-binding protein (GM2 activator) that causes GM2-gangliosidosis variant AB. FEBS Letters, 1991, 290, 1-3.	1.3	70
100	Sphingolipid biosynthesis in cultured neurons. Down-regulation of serine palmitoyltransferase by sphingoid bases. FEBS Journal, 1991, 198, 667-674.	0.2	70
101	Lipid-binding Proteins in Membrane Digestion, Antigen Presentation, and Antimicrobial Defense. Journal of Biological Chemistry, 2005, 280, 41125-41128.	1.6	70
102	Golgi-to-phagosome transport of acid sphingomyelinase and prosaposin is mediated by sortilin. Journal of Cell Science, 2010, 123, 2502-2511.	1.2	70
103	Role for LAMP-2 in endosomal cholesterol transport. Journal of Cellular and Molecular Medicine, 2011, 15, 280-295.	1.6	70
104	Saposin A Mobilizes Lipids from Low Cholesterol and High Bis(monoacylglycerol)phosphate-containing Membranes. Journal of Biological Chemistry, 2006, 281, 32451-32460.	1.6	69
105	Biosynthesis, Processing, and Targeting of Sphingolipid Activator Protein (SAP)Precursor in Cultured Human Fibroblasts. Journal of Biological Chemistry, 1996, 271, 32438-32446.	1.6	67
106	Intracellular Distribution of a Biotin-labeled Ganglioside, GM1, by Immunoelectron Microscopy After Endocytosis in Fibroblasts. Journal of Histochemistry and Cytochemistry, 1999, 47, 1005-1014.	1.3	67
107	Model for the Interaction of Membrane-Bound Substrates and Enzymes. Hydrolysis of Ganglioside GD1a by Sialidase of Neuronal Membranes Isolated from Calf Brain. FEBS Journal, 1982, 127, 245-253.	0.2	66
108	Hexosaminidase assays. Glycoconjugate Journal, 2009, 26, 945-952.	1.4	66

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109	Acid sphingomyelinase activity is regulated by membrane lipids and facilitates cholesterol transfer by NPC2. Journal of Lipid Research, 2014, 55, 2606-2619.	2.0	65
110	Occurrence of Lysoganglioside Lyso-G <sub><b>M2</b></sub> (II <sup><b>3</b></sup> -Neu5Ac-Gangliotriaosylsphingosine) in G <sub><b>M2</b></sub> Gangliosidosis Brain. Biological Chemistry Hoppe-Seyler, 1986, 367, 241-244.	1.4	64
111	Ganglioside Biosynthesis in Golgi Apparatus of Rat Liver. Stimulation by Phosphatidylglycerol and Inhibition by Tunicamycin. FEBS Journal, 1983, 134, 47-54.	0.2	63
112	Purified recombinant human prosaposin forms oligomers that bind procathepsin D and affect its autoactivation. Biochemical Journal, 2004, 383, 507-515.	1.7	63
113	The c-series gangliosides GT3, GT2 and GP1C are formed in rat liver Golgi by the same set of glycosyltransferases that catalyse the biosynthesis of asialo-, a- and b-series gangliosides. Glycobiology, 1992, 2, 137-142.	1.3	62
114	Development of an assay for the intermembrane transfer of cholesterol by Niemann-Pick C2 protein. Biological Chemistry, 2007, 388, 617-26.	1.2	60
115	Deletion of arginine (608) in acid sphingomyelinase is the prevalent mutation among Niemann-Pick disease type B patients from northern Africa. Human Genetics, 1993, 92, 325-330.	1.8	59
116	Phosphatidylinositol-3,5-Bisphosphate Is a Potent and Selective Inhibitor of Acid Sphingomyelinase. Biological Chemistry, 2003, 384, 1293-8.	1.2	59
117	The complete amino-acid sequences of human ganglioside GM2 activator protein and cerebroside sulfate activator protein. FEBS Journal, 1990, 192, 709-714.	0.2	58
118	Saposin B mobilizes lipids from cholesterol-poor and bis(monoacylglycero)phosphate-rich membranes at acidic pH. FEBS Journal, 2007, 274, 3405-3420.	2.2	58
119	Degradation of Membrane-bound Ganglioside GM2 by $\hat{l}^2$ -Hexosaminidase A. Journal of Biological Chemistry, 2001, 276, 12685-12690.	1.6	57
120	Separation and mass spectrometric characterization of covalently bound skin ceramides using LC/APCI-MS and Nano-ESI-MS/MS. Journal of Chromatography B: Analytical Technologies in the Biomedical and Life Sciences, 2007, 852, 562-570.	1.2	56
121	Molecular genetics of GM2-gangliosidosis AB variant: a novel mutation and expression in BHK cells. Human Genetics, 1993, 92, 437-440.	1.8	55
122	Ganglioside Metabolism in Health and Disease. Progress in Molecular Biology and Translational Science, 2018, 156, 1-62.	0.9	55
123	Specificity of human glucosylceramide beta-glucosidase towards synthetic glucosylsphingolipids inserted into liposomes. Kinetic studies in a detergent-free assay system. FEBS Journal, 1986, 160, 527-535.	0.2	54
124	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. Journal of Chromatography B: Analytical Technologies in the Biomedical and Life Sciences, 2009, 877, 2976-2982.	1.2	54
125	Functional Characterization of the N-glycosylation Sites of Human Acid Sphingomyelinase by Site-Directed Mutagenesis. FEBS Journal, 1997, 243, 511-517.	0.2	53
126	CLN3 Loss Disturbs Membrane Microdomain Properties and Protein Transport in Brain Endothelial Cells. Journal of Neuroscience, 2013, 33, 18065-18079.	1.7	53

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127	Mechanism of Secondary Ganglioside and Lipid Accumulation in Lysosomal Disease. International Journal of Molecular Sciences, 2020, 21, 2566.	1.8	52
128	Characterization of full-length cDNAs and the gene coding for the human GM2 activator protein. FEBS Letters, 1991, 289, 260-264.	1.3	51
129	Substrate specificity of alpha2<->3-sialyltransferases in ganglioside biosynthesis of rat liver golgi*. FEBS Journal, 1991, 195, 115-120.	0.2	51
130	Synthesis of novel NBD-GM1 and NBD-GM2 for the transfer activity of GM2-activator protein by a FRET-based assay system. Glycobiology, 2005, 15, 1302-1311.	1.3	51
131	Photoaffinity labelling of the Human GM2-activator protein. Mechanistic insight into ganglioside GM2 degradation. FEBS Journal, 2004, 271, 614-627.	0.2	49
132	Chapter 14 Sphingolipids: metabolism and cell signaling. New Comprehensive Biochemistry, 2002, 36, 373-407.	0.1	48
133	Fractionation of Primary Cultured Cerebellar Neurons: Distribution of Sialyltransferases Involved in Ganglioside Biosynthesis. Journal of Neurochemistry, 1992, 58, 1533-1537.	2.1	47
134	Metabolic and cellular bases of sphingolipidoses. Biochemical Society Transactions, 2013, 41, 1562-1568.	1.6	47
135	Molecular basis of acid sphingomyelinase dificiency in a patient with Niemann-Pick disease type A. Biochemical and Biophysical Research Communications, 1991, 179, 1187-1191.	1.0	46
136	The organization of the gene for the human cerebroside sulfate activator protein. FEBS Letters, 1991, 280, 267-270.	1.3	46
137	Amplification and overexpression of prosaposin in prostate cancer. Genes Chromosomes and Cancer, 2005, 44, 351-364.	1.5	46
138	Enzyme-linked Immunosorbent Assay for the Ganglioside GM2-Activator Protein. Screening of Normal Human Tissues and Body Fluids, of Tissues of GM2Gangliosidosis, and for Its Subcellular Localization. Hoppe-Seyler's Zeitschrift FÃ1/4r Physiologische Chemie, 1984, 365, 347-356.	1.7	45
139	Differential Regulation of Src-Family Protein Tyrosine Kinases in GPI Domains of T Lymphocyte Plasma Membranes. Biochemical and Biophysical Research Communications, 1996, 225, 801-807.	1.0	44
140	Hydrolysis of lactosylceramide by human galactosylceramidase and GM1-beta-galactosidase in a detergent-free system and its stimulation by sphingolipid activator proteins, sap-B and sap-C Activator proteins stimulate lactosylceramide hydrolysis. FEBS Journal, 1994, 222, 83-90.	0.2	43
141	Regulation of the NPC2 proteinâ€mediated cholesterol trafficking by membrane lipids. Journal of Neurochemistry, 2011, 116, 702-707.	2.1	43
142	Biosynthesis, Processing, and Intracellular Transport of GM2 Activator Protein in Human Epidermal Keratinocytes. Journal of Biological Chemistry, 1997, 272, 5199-5207.	1.6	42
143	Human acid sphingomyelinase. Assignment of the disulfide bond pattern. FEBS Journal, 2003, 270, 1076-1088.	0.2	40
144	Ceramide Synthase Schlank Is a Transcriptional Regulator Adapting Gene Expression to Energy Requirements. Cell Reports, 2018, 22, 967-978.	2.9	40

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145	Unbalanced lipolysis results in lipotoxicity and mitochondrial damage in peroxisome-deficient <i>Pex19</i> mutants. Molecular Biology of the Cell, 2018, 29, 396-407.	0.9	40
146	Sphingolipid Metabolism: Sphingoid Analogs, Sphingolipid Activator Proteins, and the Pathology of the Cella. Annals of the New York Academy of Sciences, 1998, 845, 139-151.	1.8	39
147	Accumulation of Glucosylceramide in the Absence of the Beta-Glucosidase GBA2 Alters Cytoskeletal Dynamics. PLoS Genetics, 2015, 11, e1005063.	1.5	39
148	Characterization of sphingomyelinase activity released by thrombin-stimulated platelets. Molecular and Cellular Biochemistry, 2000, 205, 75-81.	1.4	38
149	Loss of keratin 10 is accompanied by increased sebocyte proliferation and differentiation. European Journal of Cell Biology, 2004, 83, 747-759.	1.6	38
150	Postnatal Requirement of the Epithelial Sodium Channel for Maintenance of Epidermal Barrier Function. Journal of Biological Chemistry, 2008, 283, 2622-2630.	1.6	38
151	Neural precursor cell cultures from GM2 gangliosidosis animal models recapitulate the biochemical and molecular hallmarks of the brain pathology. Journal of Neurochemistry, 2009, 109, 135-147.	2.1	38
152	Lipids regulate the hydrolysis of membrane bound glucosylceramide by lysosomal $\hat{l}^2$ -glucocerebrosidase. Journal of Lipid Research, 2017, 58, 563-577.	2.0	38
153	[66] Activator proteins for lysosomal glycolipid hydrolysis. Methods in Enzymology, 1987, 138, 792-815.	0.4	37
154	Inhibitors of glycosphingolipid biosythesis. Chemical Society Reviews, 1996, 25, 371.	18.7	37
155	Sphingolipid metabolism during epidermal barrier development in mice. Journal of Lipid Research, 2002, 43, 1727-1733.	2.0	37
156	Photoaffinity Labeling of Human Lysosomal $\hat{l}^2$ -Hexosaminidase B. Journal of Biological Chemistry, 1995, 270, 23693-23699.	1.6	36
157	Characterization of two α-galactosidase mutants (Q279E and R301Q) found in an atypical variant of Fabry disease. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2000, 1501, 227-235.	1.8	36
158	Purification, Biochemical and Immunological Characterisation of Hexosaminidase A from Variant AB of Infantile GM2 Gangliosidosis. FEBS Journal, 1978, 84, 27-33.	0.2	35
159	Detection of protein mediated glycosphingolipid clustering by the use of resonance energy transfer between fluorescent labelled lipids. A method established by applying the system ganglioside GM1 and cholera toxin B subunit. Chemistry and Physics of Lipids, 1992, 62, 269-280.	1.5	35
160	Mass spectrometric analysis of neutral sphingolipids: Methods, applications, and limitations. Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids, 2011, 1811, 854-860.	1,2	35
161	My journey into the world of sphingolipids and sphingolipidoses. Proceedings of the Japan Academy Series B: Physical and Biological Sciences, 2012, 88, 554-582.	1.6	35
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