# Konrad Sandhoff

List of Publications by Year
in descending order

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| 285 | 23,048 | 6254 | 10445 |
|---|---|---|---|
| papers | citations | 80 h-index | 139 g-index |

| 303 | 303 | 303 | 16944 |
|---|---|---|---|
| all docs | docs citations | times ranked | citing authors |

| # | Article | IF | Citations |
|---|---|---|---|
| 1 | Direct observation of the nanoscale dynamics of membrane lipids in a living cell. Nature, 2009, 457, 1159-1162. | 27.8 | 1,392 |
| 2 | CD95 Signaling via Ceramide-rich Membrane Rafts. Journal of Biological Chemistry, 2001, 276, 20589-20596. | 3.4 | 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. | 7.1 | 487 |
| 4 | Acid sphingomyelinase deficient mice: a model of types A and B Niemannâ€“Pick disease. Nature Genetics, 1995, 10, 288-293. | 21.4 | 457 |
| 5 | Hsp70 stabilizes lysosomes and reverts Niemannâ€“Pick disease-associated lysosomal pathology. Nature, 2010, 463, 549-553. | 27.8 | 425 |
| 6 | Mouse models of Tayâ€“Sachs and Sandhoff diseases differ in neurologic phenotype and ganglioside metabolism. Nature Genetics, 1995, 11, 170-176. | 21.4 | 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. | 9.4 | 397 |
| 8 | Physiology and pathophysiology of sphingolipid metabolism and signaling. Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids, 2000, 1485, 63-99. | 2.4 | 372 |
| 9 | Sphingolipidsâ€”Their Metabolic Pathways and the Pathobiochemistry of Neurodegenerative Diseases. Angewandte Chemie - International Edition, 1999, 38, 1532-1568. | 13.8 | 360 |
| 10 | Gangliosides and Gangliosidoses: Principles of Molecular and Metabolic Pathogenesis. Journal of Neuroscience, 2013, 33, 10195-10208. | 3.6 | 356 |
| 11 | Apoptotic Vesicles Crossprime CD8 T Cells and Protect against Tuberculosis. Immunity, 2006, 24, 105-117. | 14.3 | 353 |
| 12 | Acidic Sphingomyelinase Mediates Entry of N. gonorrhoeae into Nonphagocytic Cells. Cell, 1997, 91, 605-615. | 28.9 | 307 |
| 13 | Sphingolipid metabolism diseases. Biochimica Et Biophysica Acta - Biomembranes, 2006, 1758, 2057-2079. | 2.6 | 306 |
| 14 | Combinatorial Ganglioside Biosynthesis. Journal of Biological Chemistry, 2002, 277, 25859-25862. | 3.4 | 271 |
| 15 | Characterization of Ceramide Synthesis. Journal of Biological Chemistry, 1997, 272, 22432-22437. | 3.4 | 266 |
| 16 | The epidermal barrier function is dependent on the serine protease CAP1/Prss8 . Journal of Cell Biology, 2005, 170, 487-496. | 5.2 | 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. | 3.4 | 245 |

| # | Article | IF | Citations |
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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. | 3.4 | 232 |
| 20 | Lysosomal degradation of membrane lipids. FEBS Letters, 2010, 584, 1700-1712. | 2.8 | 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. | 3.4 | 218 |
| 23 | Deficiency of Epidermal Protein-Bound Ï‰-Hydroxyceramides in Atopic Dermatitis. Journal of Investigative Dermatology, 2002, 119, 166-173. | 0.7 | 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. | 7.1 | 212 |
| 25 | Principles of lysosomal membrane degradation. Biochimica Et Biophysica Acta - Molecular Cell Research, 2009, 1793, 674-683. | 4.1 | 203 |
| 26 | Mice lacking both subunits of lysosomal Î²â€“hexosaminidase display gangliosidosis and mucopolysaccharidosis. Nature Genetics, 1996, 14, 348-352. | 21.4 | 194 |
| 27 | Purification, Characterization, and Biosynthesis of Human Acid Ceramidase. Journal of Biological Chemistry, 1995, 270, 11098-11102. | 3.4 | 193 |
| 28 | Acid Ceramidase Overexpression Prevents the Inhibitory Effects of Saturated Fatty Acids on Insulin Signaling. Journal of Biological Chemistry, 2005, 280, 20148-20153. | 3.4 | 188 |
| 29 | Metabolism and intracellular transport of glycosphingolipids. Biochemistry, 1990, 29, 10865-10871. | 2.5 | 178 |
| 30 | Purification and Characterization of an Activator Protein for the Degradation of Glycolipids GM2and GA2by Hexosaminidase A. Hoppe-Seyler's Zeitschrift FÃ¼r Physiologische Chemie, 1979, 360, 1837-1850. | 1.6 | 176 |
| 31 | Interactions of acid sphingomyelinase and lipid bilayers in the presence of the tricyclic antidepressant desipramine. FEBS Letters, 2004, 559, 96-98. | 2.8 | 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. | 7.1 | 169 |
| 33 | Overexpression of Acid Ceramidase Protects from Tumor Necrosis Factorâ€“Induced Cell Death. Journal of Experimental Medicine, 2000, 192, 601-612. | 8.5 | 164 |
| 34 | Topology of glycosphingolipid degradation. Trends in Cell Biology, 1996, 6, 98-103. | 7.9 | 161 |
| 35 | Incorporation of ganglioside analogs into fibroblast cell membranes. A spin-label study. Biochemistry, 1983, 22, 5041-5048. | 2.5 | 160 |
| 36 | Saposin C is required for lipid presentation by human CD1b. Nature Immunology, 2004, 5, 169-174. | 14.5 | 160 |

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| 37 | Biosynthesis and degradation of mammalian glycosphingolipids. Philosophical Transactions of the Royal Society B: Biological Sciences, 2003, 358, 847-861. | 4.0 | 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. | 2.8 | 153 |
| 40 | Nrf2 links epidermal barrier function with antioxidant defense. EMBO Molecular Medicine, 2012, 4, 364-379. | 6.9 | 153 |
| 41 | A genetic model of substrate deprivation therapy for a glycosphingolipid storage disorder. Journal of Clinical Investigation, 1999, 103, 497-505. | 8.2 | 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. | 5.5 | 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. | 2.4 | 141 |
| 46 | Lysosomal Degradation on Vesicular Membrane Surfaces. Journal of Biological Chemistry, 1998, 273, 30271-30278. | 3.4 | 140 |
| 47 | Biosynthesis of sphingolipids: Dihydroceramide and not sphinganine is desaturated by cultured cells. Biochemical and Biophysical Research Communications, 1992, 189, 14-20. | 2.1 | 139 |
| 48 | Sphingolipid Activator Proteins Are Required for Epidermal Permeability Barrier Formation. Journal of Biological Chemistry, 1999, 274, 11038-11045. | 3.4 | 138 |
| 49 | Structure of Full-Length cDNA Coding for Sulfatide Activator, a Co-Î²-Glucosidase and Two Other Homologous Proteins: Two Alternate Forms of the Sulfatide Activator1. Journal of Biochemistry, 1989, 105, 152-154. | 1.7 | 137 |
| 50 | The Reverse Activity of Human Acid Ceramidase. Journal of Biological Chemistry, 2003, 278, 29948-29953. | 3.4 | 133 |
| 51 | The Human Acid Ceramidase Gene (ASAH): Structure, Chromosomal Location, Mutation Analysis, and Expression. Genomics, 1999, 62, 223-231. | 2.9 | 130 |
| 52 | Stereoselective synthesis of Î±- C-allyl-glycopyranosides. Tetrahedron Letters, 1985, 26, 1479-1482. | 1.4 | 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. | 11.2 | 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. | 3.9 | 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. | 3.4 | 117 |
| 58 | Inhibition of Glycosphingolipid Biosynthesis Reduces Secretion of the Î²-Amyloid Precursor Protein and Amyloid Î²-Peptide*[boxs]. Journal of Biological Chemistry, 2005, 280, 28110-28117. | 3.4 | 115 |
| 59 | Systemic inflammation in glucocerebrosidase-deficient mice with minimal glucosylceramide storage. Journal of Clinical Investigation, 2002, 109, 1215-1221. | 8.2 | 114 |
| 60 | Interfacial Regulation of Acid Ceramidase Activity. Journal of Biological Chemistry, 2001, 276, 5760-5768. | 3.4 | 113 |
| 61 | Cloning and Characterization of the Full-Length cDNA and Genomic Sequences Encoding Murine Acid Ceramidase. Genomics, 1998, 50, 267-274. | 2.9 | 109 |
| 62 | The X-ray Crystal Structure of Human Î²-Hexosaminidase B Provides New Insights into Sandhoff Disease. Journal of Molecular Biology, 2003, 328, 669-681. | 4.2 | 109 |
| 63 | Biological Function of the Cellular Lipid BMPâ€"BMP as a Key Activator for Cholesterol Sorting and Membrane Digestion. Neurochemical Research, 2011, 36, 1594-1600. | 3.3 | 108 |
| 64 | Insulin receptor and lipid metabolism pathology in ataxin-2 knock-out mice. Human Molecular Genetics, 2008, 17, 1465-1481. | 2.9 | 107 |
| 65 | Anreicherung und Charakterisierung zweier Formen der menschlichen N-Acetyl-Î²-D-hexosaminidase. Hoppe-Seyler's Zeitschrift FÃ¼r Physiologische Chemie, 1971, 352, 1119-1133. | 1.6 | 105 |
| 66 | [26] Lysogangliosides: Synthesis and use in preparing labeled gangliosides. Methods in Enzymology, 1987, 138, 319-341. | 1.0 | 105 |
| 67 | Integrity and Barrier Function of the Epidermis Critically Depend on Glucosylceramide Synthesis. Journal of Biological Chemistry, 2007, 282, 3083-3094. | 3.4 | 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. | 2.9 | 104 |
| 70 | Human Acid Ceramidase. Journal of Biological Chemistry, 2001, 276, 35352-35360. | 3.4 | 98 |
| 71 | Emerging mechanisms of drug-induced phospholipidosis. Biological Chemistry, 2019, 401, 31-46. | 2.5 | 97 |
| 72 | PAR2 absence completely rescues inflammation and ichthyosis caused by altered CAP1/Prss8 expression in mouse skin. Nature Communications, 2011, 2, 161. | 12.8 | 96 |

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| 73 | Stimulation of Acid Sphingomyelinase Activity by Lysosomal Lipids and Sphingolipid Activator Proteins. Biological Chemistry, 2001, 382, 283-90. | 2.5 | 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 FÃ¼r Physiologische Chemie, 1977, 358, 779-788. | 1.6 | 89 |
| 75 | Purification and Characterization of Recombinant, Human Acid Ceramidase. Journal of Biological Chemistry, 2003, 278, 32978-32986. | 3.4 | 88 |
| 76 | Sphingolipids and lysosomal pathologies. Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids, 2014, 1841, 799-810. | 2.4 | 88 |
| 77 | Normal epidermal differentiation but impaired skin-barrier formation upon keratinocyte-restricted IKK1 ablation. Nature Cell Biology, 2007, 9, 461-469. | 10.3 | 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 proteinâ€bound epidermal glucosylceramides in Î²â€glucocerebrosidase deficient type 2 Gaucher mice. FEBS Letters, 1999, 447, 167-170. | 2.8 | 86 |
| 80 | Sphingolipid Storage Affects Autophagic Metabolism of the Amyloid Precursor Protein and Promotes Aβ Generation. Journal of Neuroscience, 2011, 31, 1837-1849. | 3.6 | 82 |
| 81 | Physiological Substrates for Human Lysosomal β-Hexosaminidase S. Journal of Biological Chemistry, 2002, 277, 2562-2572. | 3.4 | 81 |
| 82 | Activation of Nrf2 in keratinocytes causes chloracne (MADISH)â€like skin disease in mice. EMBO Molecular Medicine, 2014, 6, 442-457. | 6.9 | 81 |
| 83 | Degradation of Membrane-bound Ganglioside GM1. Journal of Biological Chemistry, 2000, 275, 35814-35819. | 3.4 | 79 |
| 84 | Killing from the inside. Nature, 2013, 502, 312-313. | 27.8 | 79 |
| 85 | Crystal Structures of Human Saposins C and D: Implications for Lipid Recognition and Membrane Interactions. Structure, 2008, 16, 809-817. | 3.3 | 78 |
| 86 | Lysosomal Glycosphingolipid Storage Diseases. Annual Review of Biochemistry, 2019, 88, 461-485. | 11.1 | 78 |
| 87 | Molecular analysis of acid ceramidase deficiency in patients with Farber disease. Human Mutation, 2001, 17, 199-209. | 2.5 | 76 |
| 88 | Schlank, a member of the ceramide synthase family controls growth and body fat in Drosophila. EMBO Journal, 2009, 28, 3706-3716. | 7.8 | 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. | 4.1 | 73 |

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| 91 | Systemic inflammation in glucocerebrosidase-deficient mice with minimal glucosylceramide storage. Journal of Clinical Investigation, 2002, 109, 1215-1221. | 8.2 | 73 |
| 92 | Genomic Organization of the Gene Coding for TIRC7, a Novel Membrane Protein Essential for T Cell Activation. Genomics, 1999, 57, 398-406. | 2.9 | 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. | 2.8 | 72 |
| 95 | Pyrene-labeled gangliosides: micelle formation in aqueous solution, lateral diffusion, and thermotropic behavior in phosphatidylcholine bilayers. Biochemistry, 1987, 26, 5943-5952. | 2.5 | 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. | 3.4 | 71 |
| 97 | Purification and Characterization of a Magnesium-dependent Neutral Sphingomyelinase from Bovine Brain. Journal of Biological Chemistry, 2000, 275, 7641-7647. | 3.4 | 71 |
| 98 | Role of endosomal membrane lipids and NPC2 in cholesterol transfer and membrane fusion. Journal of Lipid Research, 2010, 51, 1747-1760. | 4.2 | 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. | 2.8 | 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. | 3.4 | 70 |
| 102 | Golgi-to-phagosome transport of acid sphingomyelinase and prosaposin is mediated by sortilin. Journal of Cell Science, 2010, 123, 2502-2511. | 2.0 | 70 |
| 103 | Role for LAMP-2 in endosomal cholesterol transport. Journal of Cellular and Molecular Medicine, 2011, 15, 280-295. | 3.6 | 70 |
| 104 | Saposin A Mobilizes Lipids from Low Cholesterol and High Bis(monoacylglycerol)phosphate-containing Membranes. Journal of Biological Chemistry, 2006, 281, 32451-32460. | 3.4 | 69 |
| 105 | Biosynthesis, Processing, and Targeting of Sphingolipid Activator Protein (SAP)Precursor in Cultured Human Fibroblasts. Journal of Biological Chemistry, 1996, 271, 32438-32446. | 3.4 | 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. | 2.5 | 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. | 2.7 | 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. | 4.2 | 65 |
| 110 | Occurrence of Lysoganglioside Lyso-G<sub>M2</sub>(II<sup>3</sup>-Neu5Ac-Gangliotriaosylsphingosine) in G<sub>M2</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. | 3.7 | 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. | 2.5 | 62 |
| 114 | Development of an assay for the intermembrane transfer of cholesterol by Niemann-Pick C2 protein. Biological Chemistry, 2007, 388, 617-26. | 2.5 | 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. | 3.8 | 59 |
| 116 | Phosphatidylinositol-3,5-Bisphosphate Is a Potent and Selective Inhibitor of Acid Sphingomyelinase. Biological Chemistry, 2003, 384, 1293-8. | 2.5 | 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 | Saposinâ€ƒB mobilizes lipids from cholesterol-poor and bis(monoacylglycero)phosphate-rich membranes at acidic pH. FEBS Journal, 2007, 274, 3405-3420. | 4.7 | 58 |
| 119 | Degradation of Membrane-bound Ganglioside GM2 by Î²-Hexosaminidase A. Journal of Biological Chemistry, 2001, 276, 12685-12690. | 3.4 | 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. | 2.3 | 56 |
| 121 | Molecular genetics of GM2-gangliosidosis AB variant: a novel mutation and expression in BHK cells. Human Genetics, 1993, 92, 437-440. | 3.8 | 55 |
| 122 | Ganglioside Metabolism in Health and Disease. Progress in Molecular Biology and Translational Science, 2018, 156, 1-62. | 1.7 | 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. | 2.3 | 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. | 3.6 | 53 |

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| 127 | Mechanism of Secondary Ganglioside and Lipid Accumulation in Lysosomal Disease. International Journal of Molecular Sciences, 2020, 21, 2566. | 4.1 | 52 |
| 128 | Characterization of full-length cDNAs and the gene coding for the human GM2 activator protein. FEBS Letters, 1991, 289, 260-264. | 2.8 | 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. | 2.5 | 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. | 3.9 | 47 |
| 134 | Metabolic and cellular bases of sphingolipidoses. Biochemical Society Transactions, 2013, 41, 1562-1568. | 3.4 | 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. | 2.1 | 46 |
| 136 | The organization of the gene for the human cerebroside sulfate activator protein. FEBS Letters, 1991, 280, 267-270. | 2.8 | 46 |
| 137 | Amplification and overexpression of prosaposin in prostate cancer. Genes Chromosomes and Cancer, 2005, 44, 351-364. | 2.8 | 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 FÃ¼r Physiologische Chemie, 1984, 365, 347-356. | 1.6 | 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. | 2.1 | 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. | 3.9 | 43 |
| 142 | Biosynthesis, Processing, and Intracellular Transport of GM2 Activator Protein in Human Epidermal Keratinocytes. Journal of Biological Chemistry, 1997, 272, 5199-5207. | 3.4 | 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. | 6.4 | 40 |

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| 145 | Unbalanced lipolysis results in lipotoxicity and mitochondrial damage in peroxisome-deficient Pex19 mutants. Molecular Biology of the Cell, 2018, 29, 396-407. | 2.1 | 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. | 3.8 | 39 |
| 147 | Accumulation of Glucosylceramide in the Absence of the Beta-Glucosidase GBA2 Alters Cytoskeletal Dynamics. PLoS Genetics, 2015, 11, e1005063. | 3.5 | 39 |
| 148 | Characterization of sphingomyelinase activity released by thrombin-stimulated platelets. Molecular and Cellular Biochemistry, 2000, 205, 75-81. | 3.1 | 38 |
| 149 | Loss of keratin 10 is accompanied by increased sebocyte proliferation and differentiation. European Journal of Cell Biology, 2004, 83, 747-759. | 3.6 | 38 |
| 150 | Postnatal Requirement of the Epithelial Sodium Channel for Maintenance of Epidermal Barrier Function. Journal of Biological Chemistry, 2008, 283, 2622-2630. | 3.4 | 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. | 3.9 | 38 |
| 152 | Lipids regulate the hydrolysis of membrane bound glucosylceramide by lysosomal Î²-glucocerebrosidase. Journal of Lipid Research, 2017, 58, 563-577. | 4.2 | 38 |
| 153 | [66] Activator proteins for lysosomal glycolipid hydrolysis. Methods in Enzymology, 1987, 138, 792-815. | 1.0 | 37 |
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