Edward H Schuchman

List of Publications by Year
in descending order

160 papers	13,600 citations	19657 61 h-index	22832 112 g-index
165 all docs	165 docs citations	165 times ranked	11354 citing authors

#	Article	IF	Citations
1	Acid Sphingomyelinaseâ€“Deficient Human Lymphoblasts and Mice Are Defective in Radiation-Induced Apoptosis. Cell, 1996, 86, 189-199.	28.9	780
2	Niemann-Pick disease type C1 is a sphingosine storage disease that causes deregulation of lysosomal calcium. Nature Medicine, 2008, 14, 1247-1255.	30.7	730
3	Oocyte apoptosis is suppressed by disruption of the acid sphingomyelinase gene or by sphingosine -1-phosphate therapy. Nature Medicine, 2000, 6, 1109-1114.	30.7	552
4	Control of Endothelial Targeting and Intracellular Delivery of Therapeutic Enzymes by Modulating the Size and Shape of ICAM-1-targeted Carriers. Molecular Therapy, 2008, 16, 1450-1458.	8.2	506
5	Acid sphingomyelinase activity triggers microparticle release from glial cells. EMBO Journal, 2009, 28, 1043-1054.	7.8	499
6	Acid sphingomyelinase deficient mice: a model of types A and B Niemannâ€“Pick disease. Nature Genetics, 1995, 10, 288-293.	21.4	457
7	Deregulation of sphingolipid metabolism in Alzheimer's disease. Neurobiology of Aging, 2010, 31, 398-408.	3.1	447
8	Lipopolysaccharide Induces Disseminated Endothelial Apoptosis Requiring Ceramide Generation. Journal of Experimental Medicine, 1997, 186, 1831-1841.	8.5	412
9	Exocytosis of acid sphingomyelinase by wounded cells promotes endocytosis and plasma membrane repair. Journal of Cell Biology, 2010, 189, 1027-1038.	5.2	301
10	Zn2+-stimulated Sphingomyelinase Is Secreted by Many Cell Types and Is a Product of the Acid Sphingomyelinase Gene. Journal of Biological Chemistry, 1996, 271, 18431-18436.	3.4	257
11	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
12	The Cellular Trafficking and Zinc Dependence of Secretory and Lysosomal Sphingomyelinase, Two Products of the Acid Sphingomyelinase Gene. Journal of Biological Chemistry, 1998, 273, 18250-18259.	3.4	219
13	Types A and B Niemann-Pick disease. Best Practice and Research in Clinical Endocrinology and Metabolism, 2015, 29, 237-247.	4.7	200
14	Types A and B Niemann-Pick disease. Molecular Genetics and Metabolism, 2017, 120, 27-33.	1.1	196
15	Purification, Characterization, and Biosynthesis of Human Acid Ceramidase. Journal of Biological Chemistry, 1995, 270, 11098-11102.	3.4	193
16	The unexpected role of acid sphingomyelinase in cell death and the pathophysiology of common diseases. FASEB Journal, 2008, 22, 3419-3431.	0.5	189
17	Pivotal Role for Acidic Sphingomyelinase in Cerebral Ischemia-Induced Ceramide and Cytokine Production, and Neuronal Apoptosis. Journal of Molecular Neuroscience, 2000, 15, 85-98.	2.3	188
18	Mechanism of Glycosaminoglycan-Mediated Bone and Joint Disease. American Journal of Pathology, 2008, 172, 112-122.	3.8	188

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19	Acid ceramidase and human disease. Biochimica Et Biophysica Acta - Biomembranes, 2006, 1758, 2133-2138.	2.6	171
20	Involvement of the Toll-like receptor 4 pathway and use of TNF-Î± antagonists for treatment of the mucopolysaccharidoses. Proceedings of the National Academy of Sciences of the United States of America, 2010, 107, 222-227.	7.1	155
21	The Natural History of Type B Niemann-Pick Disease: Results From a 10-Year Longitudinal Study. Pediatrics, 2004, 114, e672-e677.	2.1	138
22	Role of Acidic Sphingomyelinase in Fas/CD95-mediated Cell Death. Journal of Biological Chemistry, 2000, 275, 8657-8663.	3.4	137
23	Acid Sphingomyelinase Promotes Lipoprotein Retention Within Early Atheromata and Accelerates Lesion Progression. Arteriosclerosis, Thrombosis, and Vascular Biology, 2008, 28, 1723-1730.	2.4	137
24	The Demographics and Distribution of Type B Niemann-Pick Disease: Novel Mutations Lead to New Genotype/Phenotype Correlations. American Journal of Human Genetics, 2002, 71, 1413-1419.	6.2	136
25	Structural organization and complete nucleotide sequence of the gene encoding human acid sphingomyelinase (SMPD1). Genomics, 1992, 12, 197-205.	2.9	134
26	The Reverse Activity of Human Acid Ceramidase. Journal of Biological Chemistry, 2003, 278, 29948-29953.	3.4	133
27	The Human Acid Ceramidase Gene (ASAH): Structure, Chromosomal Location, Mutation Analysis, and Expression. Genomics, 1999, 62, 223-231.	2.9	130
28	Articular Chondrocytes from Animals with a Dermatan Sulfate Storage Disease Undergo a High Rate of Apoptosis and Release Nitric Oxide and Inflammatory Cytokines: A Possible Mechanism Underlying Degenerative Joint Disease in the Mucopolysaccharidoses. Laboratory Investigation, 2001, 81, 1319-1328.	3.7	130
29	Acid sphingomyelinase modulates the autophagic process by controlling lysosomal biogenesis in Alzheimerâ€™s disease. Journal of Experimental Medicine, 2014, 211, 1551-1570.	8.5	128
30	Infusion of recombinant human acid sphingomyelinase into Niemannâ€Pick disease mice leads to visceral, but not neurological, correction of the pathophysiology. FASEB Journal, 2000, 14, 1988-1995.	0.5	126
31	Acid sphingomyelinase, cell membranes and human disease: Lessons from Niemannâ€“Pick disease. FEBS Letters, 2010, 584, 1895-1900.	2.8	117
32	Interfacial Regulation of Acid Ceramidase Activity. Journal of Biological Chemistry, 2001, 276, 5760-5768.	3.4	113
33	Lipid abnormalities in children with types A and B Niemann Pick disease. Journal of Pediatrics, 2004, 145, 77-81.	1.8	113
34	Lysosomal enzyme delivery by ICAM-1-targeted nanocarriers bypassing glycosylation- and clathrin-dependent endocytosis. Molecular Therapy, 2006, 13, 135-141.	8.2	113
35	Cloning and Characterization of the Full-Length cDNA and Genomic Sequences Encoding Murine Acid Ceramidase. Genomics, 1998, 50, 267-274.	2.9	109
36	Sphingoid long chain bases prevent lung infection by Pseudomonas aeruginosa. EMBO Molecular Medicine, 2014, 6, 1205-1214.	6.9	109

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37	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
38	Morbidity and mortality in type B Niemannâ€“Pick disease. Genetics in Medicine, 2013, 15, 618-623.	2.4	99
39	Human Acid Ceramidase. Journal of Biological Chemistry, 2001, 276, 35352-35360.	3.4	98
40	Acid ceramidase is a novel factor required for early embryo survival. FASEB Journal, 2007, 21, 1403-1409.	0.5	97
41	Delivery of Acid Sphingomyelinase in Normal and Niemann-Pick Disease Mice Using Intercellular Adhesion Molecule-1-Targeted Polymer Nanocarriers. Journal of Pharmacology and Experimental Therapeutics, 2008, 325, 400-408.	2.5	97
42	Systemic ceramide accumulation leads to severe and varied pathological consequences. EMBO Molecular Medicine, 2013, 5, 827-842.	6.9	90
43	Purification and Characterization of Recombinant, Human Acid Ceramidase. Journal of Biological Chemistry, 2003, 278, 32978-32986.	3.4	88
44	Characterization of human acid sphingomyelinase purified from the media of overexpressing Chinese hamster ovary cells. BBA - Proteins and Proteomics, 1999, 1432, 251-264.	2.1	87
45	Î²1-Integrin Accumulates in Cystic Fibrosis Luminal Airway Epithelial Membranes and Decreases Sphingosine, Promoting Bacterial Infections. Cell Host and Microbe, 2017, 21, 707-718.e8.	11.0	86
46	AAV Vector-Mediated Correction of Brain Pathology in a Mouse Model of Niemannâ€“Pick A Disease. Molecular Therapy, 2005, 11, 754-762.	8.2	85
47	Patterned cerebellar Purkinje cell death in a transgenic mouse model of Niemann Pick type A/B disease. European Journal of Neuroscience, 2001, 13, 1873-1880.	2.6	82
48	Pentosan Polysulfate: A Novel Therapy for the Mucopolysaccharidoses. PLoS ONE, 2013, 8, e54459.	2.5	82
49	Gene transfer of human acid sphingomyelinase corrects neuropathology and motor deficits in a mouse model of Niemann-Pick type A disease. Proceedings of the National Academy of Sciences of the United States of America, 2005, 102, 17822-17827.	7.1	78
50	Ceramide Is Upregulated and Associated With Mortality in Patients With Chronic Heart Failure. Canadian Journal of Cardiology, 2015, 31, 357-363.	1.7	78
51	Consensus recommendation for a diagnostic guideline for acid sphingomyelinase deficiency. Genetics in Medicine, 2017, 19, 967-974.	2.4	77
52	Regional assignment of the human acid sphingomyelinase gene (SMPD1) by PCR analysis of somatic cell hybrids and in situ hybridization to 11p15.1â†'p15.4. Genomics, 1991, 9, 229-234.	2.9	76
53	Molecular analysis of acid ceramidase deficiency in patients with Farber disease. Human Mutation, 2001, 17, 199-209.	2.5	76
54	Acid ceramidase is upregulated in AML and represents a novel therapeutic target. Oncotarget, 2016, 7, 83208-83222.	1.8	73

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55	Reproductive Pathology and Sperm Physiology in Acid Sphingomyelinase-Deficient Mice. American Journal of Pathology, 2002, 161, 1061-1075.	3.8	68
56	Improvement in Lipid and Protein Trafficking in Niemann-Pick C1 Cells by Correction of a Secondary Enzyme Defect. Traffic, 2010, 11, 601-615.	2.7	68
57	Neuronal SphK1 acetylates COX2 and contributes to pathogenesis in a model of Alzheimerâ€™s Disease. Nature Communications, 2018, 9, 1479.	12.8	68
58	Analysis of the Lung Pathology and Alveolar Macrophage Function in the Acid Sphingomyelinaseâ€“Deficient Mouse Model of Niemann-Pick Disease. Laboratory Investigation, 2001, 81, 987-999.	3.7	66
59	Intracerebroventricular infusion of acid sphingomyelinase corrects CNS manifestations in a mouse model of Niemannâ€“Pick A disease. Experimental Neurology, 2009, 215, 349-357.	4.1	66
60	Elevation of ceramide and activation of secretory acid sphingomyelinase in patients with acute coronary syndromes. Coronary Artery Disease, 2014, 25, 230-235.	0.7	66
61	Combination brain and systemic injections of AAV provide maximal functional and survival benefits in the Niemann-Pick mouse. Proceedings of the National Academy of Sciences of the United States of America, 2007, 104, 9505-9510.	7.1	65
62	Imprinting at the SMPD1 Locus: Implications for Acid Sphingomyelinaseâ€“Deficient Niemann-Pick Disease. American Journal of Human Genetics, 2006, 78, 865-870.	6.2	64
63	Autoproteolytic Cleavage and Activation of Human Acid Ceramidase. Journal of Biological Chemistry, 2008, 283, 11253-11259.	3.4	63
64	Simultaneous quantitative analysis of ceramide and sphingosine in mouse blood by naphthalene-2,3-dicarboxyaldehyde derivatization after hydrolysis with ceramidase. Analytical Biochemistry, 2005, 340, 113-122.	2.4	62
65	Acid Sphingomyelinase Deficiency Attenuates Bleomycin-Induced Lung Inflammation and Fibrosis in Mice. Cellular Physiology and Biochemistry, 2010, 26, 749-760.	1.6	61
66	Brain pathology in Niemann Pick disease type A: insights from the acid sphingomyelinase knockout mice. Journal of Neurochemistry, 2011, 116, 779-788.	3.9	61
67	Pathological roles of the VEGF/SphK pathway in Niemannâ€“Pick type C neurons. Nature Communications, 2014, 5, 5514.	12.8	61
68	Growth restriction in children with type B Niemann-Pick disease. Journal of Pediatrics, 2003, 142, 424-428.	1.8	59
69	A Fluorescence-Based, High-Throughput Sphingomyelin Assay for the Analysis of Niemannâ€“Pick Disease and Other Disorders of Sphingomyelin Metabolism. Analytical Biochemistry, 2002, 306, 115-123.	2.4	58
70	Recommendations for clinical monitoring of patients with acid sphingomyelinase deficiency (ASMD). Molecular Genetics and Metabolism, 2019, 126, 98-105.	1.1	56
71	A fluorescence-based, high-performance liquid chromatographic assay to determine acid sphingomyelinase activity and diagnose types A and B Niemannâ€“Pick disease. Analytical Biochemistry, 2003, 314, 116-120.	2.4	55
72	Identification and characterization ofSMPD1mutations causing Niemann-Pick types A and B in Spanish patients. Human Mutation, 2009, 30, 1117-1122.	2.5	54

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73	Functional Characterization of the N-glycosylation Sites of Human Acid Sphingomyelinase by Site-Directed Mutagenesis. FEBS Journal, 1997, 243, 511-517.	0.2	53
74	Lipid content of brain, brain membrane lipid domains, and neurons from acid sphingomyelinase deficient mice. Journal of Neurochemistry, 2008, 107, 329-338.	3.9	53
75	Neuropeptide Y regulates the hematopoietic stem cell microenvironment and prevents nerve injury in the bone marrow. EMBO Journal, 2015, 34, 1648-1660.	7.8	53
76	Characterization of common SMPD1 mutations causing types A and B Niemann-Pick disease and generation of mutation-specific mouse models. Molecular Genetics and Metabolism, 2008, 95, 152-162.	1.1	49
77	Comparative binding, endocytosis, and biodistribution of antibodies and antibodyâ€coated carriers for targeted delivery of lysosomal enzymes to ICAMâ€1 versus transferrin receptor. Journal of Inherited Metabolic Disease, 2013, 36, 467-477.	3.6	49
78	AAV8-Mediated Hepatic Expression of Acid Sphingomyelinase Corrects the Metabolic Defect in the Visceral Organs of a Mouse Model of Niemannâ€“Pick Disease. Molecular Therapy, 2005, 12, 431-440.	8.2	46
79	Neuropathology of the acid sphingomyelinase knockout mouse model of Niemann-Pick A disease including structureâ€“function studies associated with cerebellar Purkinje cell degeneration. Experimental Neurology, 2008, 214, 181-192.	4.1	45
80	Type A Niemann-Pick disease: A frameshift mutation in the acid sphingomyelinase gene (fsP330) occurs in Ashkenazi Jewish patients. Human Mutation, 1993, 2, 317-319.	2.5	44
81	Identification and Characterization of Eight Novel SMPD1 Mutations Causing Types A and B Niemann-Pick Disease. Molecular Medicine, 2010, 16, 316-321.	4.4	44
82	Alveolar lipoproteinosis in an acid sphingomyelinase-deficient mouse model of Niemann-Pick disease. American Journal of Physiology - Lung Cellular and Molecular Physiology, 2003, 284, L518-L525.	2.9	43
83	Pharmacological reversion of sphingomyelinâ€induced dendritic spine anomalies in a Niemann Pick disease type <scp>A</scp> mouse model. EMBO Molecular Medicine, 2014, 6, 398-413.	6.9	42
84	Acid Sphingomyelinase Overexpression Enhances the Antineoplastic Effects of Irradiation In Vitro and In Vivo. Molecular Therapy, 2008, 16, 1565-1571.	8.2	41
85	InÂvivo performance of polymer nanocarriers dually-targeted to epitopes of the same or different receptors. Biomaterials, 2013, 34, 3459-3466.	11.4	41
86	Dose Responsive Effects of Subcutaneous Pentosan Polysulfate Injection in Mucopolysaccharidosis Type VI Rats and Comparison to Oral Treatment. PLoS ONE, 2014, 9, e100882.	2.5	40
87	Construction of Conditional Acid Ceramidase Knockout Mice andin vivoEffects on Oocyte Development and Fertility. Cellular Physiology and Biochemistry, 2012, 30, 735-748.	1.6	39
88	Vascular and Neurogenic Rejuvenation in Aging Mice by Modulation of ASM. Neuron, 2018, 100, 167-182.e9.	8.1	39
89	Bone Marrow Transplantation in Acid Sphingomyelinase-Deficient Mice: Engraftment and Cell Migration Into the Brain as a Function of Radiation, Age, and Phenotype. Blood, 1997, 90, 444-452.	1.4	38
90	Acid ceramidase and the treatment of ceramide diseases: The expanding role of enzyme replacement therapy. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2016, 1862, 1459-1471.	3.8	38

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91	Pentosan Polysulfate: Oral Versus Subcutaneous Injection in Mucopolysaccharidosis Type I Dogs. PLoS ONE, 2016, 11, e0153136.	2.5	36
92	Enzyme replacement therapy for Farber disease: Proof-of-concept studies in cells and mice. BBA Clinical, 2017, 7, 85-96.	4.1	36
93	Adeno-associated viral vector serotype 9â€“based gene therapy for Niemann-Pick disease type A. Science Translational Medicine, 2019, 11, .	12.4	36
94	BIOCHEMICAL, PATHOLOGICAL, AND CLINICAL RESPONSE TO TRANSPLANTATION OF NORMAL BONE MARROW CELLS INTO ACID SPHINGOMYELINASE-DEFICIENT MICE1. Transplantation, 1998, 65, 884-892.	1.0	36
95	Acid Ceramidase Deficiency is characterized by a unique plasma cytokine and ceramide profile that is altered by therapy. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2017, 1863, 386-394.	3.8	35
96	Control of CD1d-restricted antigen presentation and inflammation by sphingomyelin. Nature Immunology, 2019, 20, 1644-1655.	14.5	35
97	Neuropeptide Y Induces Hematopoietic Stem/Progenitor Cell Mobilization by Regulating Matrix Metalloproteinase-9 Activity Through Y1 Receptor in Osteoblasts. Stem Cells, 2016, 34, 2145-2156.	3.2	33
98	Identification of Novel Biomarkers for Niemannâ€“Pick Disease Using Gene Expression Analysis of Acid Sphingomyelinase Knockout Mice. Molecular Therapy, 2006, 13, 556-564.	8.2	32
99	A Fluorescence-Based High-Performance Liquid Chromatographic Assay to Determine Acid Ceramidase Activity. Analytical Biochemistry, 1999, 274, 264-269.	2.4	31
100	Secondary Alterations of Sphingolipid Metabolism in Lysosomal Storage Diseases. Neurochemical Research, 2011, 36, 1654-1668.	3.3	31
101	N-AS-triggered SPMs are direct regulators of microglia in a model of Alzheimerâ€™s disease. Nature Communications, 2020, 11, 2358.	12.8	31
102	Identification of a missense mutation (S436R) in the acid sphingomyelinase gene from a Japanese patient with type B Niemann-Pick disease. Human Mutation, 1992, 1, 70-71.	2.5	30
103	Alterations of myelinâ€specific proteins and sphingolipids characterize the brains of acid sphingomyelinaseâ€deficient mice, an animal model of Niemannâ€“Pick disease type A. Journal of Neurochemistry, 2009, 109, 105-115.	3.9	30
104	Acid Sphingomyelinase Mediates Oxidized-LDL Induced Apoptosis in Macrophage <i>via</i> Endoplasmic Reticulum Stress. Journal of Atherosclerosis and Thrombosis, 2016, 23, 1111-1125.	2.0	30
105	Acid ceramidase improves the quality of oocytes and embryos and the outcome of in vitro fertilization. FASEB Journal, 2010, 24, 1229-1238.	0.5	28
106	Ceramide and Ischemia/Reperfusion Injury. Journal of Lipids, 2018, 2018, 1-11.	4.8	28
107	Merits of Combination Cortical, Subcortical, and Cerebellar Injections for the Treatment of Niemann-Pick Disease Type A. Molecular Therapy, 2012, 20, 1893-1901.	8.2	27
108	Enhanced Delivery and Effects of Acid Sphingomyelinase by ICAM-1-Targeted Nanocarriers in Type B Niemann-Pick Disease Mice. Molecular Therapy, 2017, 25, 1686-1696.	8.2	27

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109	Recombinant Acid Ceramidase Reduces Inflammation and Infection in Cystic Fibrosis. American Journal of Respiratory and Critical Care Medicine, 2020, 202, 1133-1145.	5.6	26
110	Ceramides are necessary and sufficient for diet-induced impairment of thermogenic adipocytes. Molecular Metabolism, 2021, 45, 101145.	6.5	26
111	A novel polymorphism in the human acid sphingomyelinase gene due to size variation of the signal peptide region. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 1995, 1270, 207-210.	3.8	25
112	A lipid analogue that inhibits sphingomyelin hydrolysis and synthesis, increases ceramide, and leads to cell death. Journal of Lipid Research, 2005, 46, 2315-2324.	4.2	25
113	Quantitative analysis of sphingosine-1-phosphate by HPLC after napthalene-2,3-dicarboxaldehyde (NDA) derivatization. Journal of Chromatography B: Analytical Technologies in the Biomedical and Life Sciences, 2009, 877, 983-990.	2.3	25
114	The molecular medicine of acid ceramidase. Biological Chemistry, 2015, 396, 759-765.	2.5	25
115	Sphingolipids as targets for inhalation treatment of cystic fibrosis. Advanced Drug Delivery Reviews, 2018, 133, 66-75.	13.7	25
116	Recombinant Human Acid Sphingomyelinase as an Adjuvant to Sorafenib Treatment of Experimental Liver Cancer. PLoS ONE, 2013, 8, e65620.	2.5	25
117	Fluorescence-Based Selection of Gene-Corrected Hematopoietic Stem and Progenitor Cells From Acid Sphingomyelinase-Deficient Mice: Implications for Niemann-Pick Disease Gene Therapy and the Development of Improved Stem Cell Gene Transfer Procedures. Blood, 1999, 93, 80-86.	1.4	24
118	Quantitative Systems Pharmacology Modeling of Acid Sphingomyelinase Deficiency and the Enzyme Replacement Therapy Olipudase Alfa Is an Innovative Tool for Linking Pathophysiology and Pharmacology. CPT: Pharmacometrics and Systems Pharmacology, 2018, 7, 442-452.	2.5	24
119	Sperm Abnormalities in Heterozygous Acid Sphingomyelinase Knockout Mice Reveal a Novel Approach for the Prevention of Genetic Diseases. American Journal of Pathology, 2007, 170, 2077-2088.	3.8	23
120	Use of Acid Sphingomyelinase for Cancer Therapy. Advances in Cancer Research, 2013, 117, 91-115.	5.0	23
121	Loss of acid ceramidase in myeloid cells suppresses intestinal neutrophil recruitment. FASEB Journal, 2018, 32, 2339-2353.	0.5	22
122	Epidemiological, clinical and biochemical characterization of the p.(Ala359Asp) SMPD1 variant causing Niemannâ€“Pick disease type B. European Journal of Human Genetics, 2016, 24, 208-213.	2.8	20
123	An Enzymatic Assay for Quantifying Sphingomyelin in Tissues and Plasma from Humans and Mice with Niemannâ€“Pick Disease. Analytical Biochemistry, 2001, 293, 204-211.	2.4	19
124	Identification of a 3â€² acceptor splice site mutation (g2610c) in the acid sphingomyelinase gene of patients with Niemann - Pick disease. Human Molecular Genetics, 1993, 2, 205-206.	2.9	18
125	Two new mutations in the acid sphingomyelinase gene causing type a Niemann-Pick disease: N389T and R441X. Human Mutation, 1995, 6, 352-354.	2.5	18
126	Acid Ceramidase Maintains the Chondrogenic Phenotype of Expanded Primary Chondrocytes and Improves the Chondrogenic Differentiation of Bone Marrow-Derived Mesenchymal Stem Cells. PLoS ONE, 2013, 8, e62715.	2.5	18

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127	KLF6 is one transcription factor involved in regulating acid ceramidase gene expression. Biochimica Et Biophysica Acta Gene Regulatory Mechanisms, 2005, 1732, 82-87.	2.4	17
128	Activity-Based Imaging of Acid Ceramidase in Living Cells. Journal of the American Chemical Society, 2019, 141, 7736-7742.	13.7	17
129	Mouse Models of Niemann-Pick Disease: Mutation Analysis and Chromosomal Mapping Rule Out the Type A and B Forms. Genomics, 1993, 18, 450-451.	2.9	16
130	CD40 Enhances Sphingolipids in Orbital Fibroblasts: Potential Role of Sphingosine-1-Phosphate in Inflammatory T-Cell Migration in Graves' Orbitopathy. , 2018, 59, 5391.		16
131	Multi-omic profiles of hepatic metabolism in TPN-fed preterm pigs administered new generation lipid emulsions. Journal of Lipid Research, 2016, 57, 1696-1711.	4.2	15
132	Polyarticular Arthritis and Spinal Muscular Atrophy in Acid Ceramidase Deficiency. Pediatrics, 2016, 138, .	2.1	15
133	Identification of Cystatin SA as a Novel Inhibitor of Acid Ceramidase. Journal of Biological Chemistry, 2011, 286, 35624-35633.	3.4	13
134	Pentosan Polysulfate Treatment of Mucopolysaccharidosis Type IIIA Mice. JIMD Reports, 2018, 43, 37-52.	1.5	13
135	Inhibition of fatty acid amide hydrolase prevents pathology in neurovisceral acid sphingomyelinase deficiency by rescuing defective endocannabinoid signaling. EMBO Molecular Medicine, 2020, 12, e11776.	6.9	13
136	Discovery of a dual-action small molecule that improves neuropathological features of Alzheimerâ€™s disease mice. Proceedings of the National Academy of Sciences of the United States of America, 2022, 119, .	7.1	12
137	Pulmonary delivery of recombinant acid sphingomyelinase improves clearance of lysosomal sphingomyelin from the lungs of a murine model of Niemannâ€“Pick disease. Molecular Genetics and Metabolism, 2009, 97, 35-42.	1.1	10
138	Comparative Effects of Recombinant Acid Sphingomyelinase Administration by Different Routes in Niemann-Pick Disease Mice. Experimental Animals, 2004, 53, 417-421.	1.1	8
139	Safety Study of Sodium Pentosan Polysulfate for Adult Patients with Mucopolysaccharidosis Type II. Diagnostics, 2019, 9, 226.	2.6	8
140	Acid Ceramidase Protects Against Hepatic Ischemia/Reperfusion Injury by Modulating Sphingolipid Metabolism and Reducing Inflammation and Oxidative Stress. Frontiers in Cell and Developmental Biology, 2021, 9, 633657.	3.7	8
141	Potential role of acid sphingomyelinase in environmental health. Journal of Central South University (Medical Sciences), 2012, 37, 109-25.	0.1	8
142	Gene therapy for genetic diseases. Pediatrics International, 1998, 40, 191-203.	0.5	7
143	Gene therapy for neurodegenerative diseases: fact or fiction?. British Journal of Psychiatry, 2001, 178, 392-394.	2.8	7
144	Characterization of the Subventricular-Thalamo-Cortical Circuit in the NP-C Mouse Brain, and New Insights Regarding Treatment. Molecular Therapy, 2019, 27, 1507-1526.	8.2	7

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145	Apolipoprotein D-mediated preservation of lysosomal function promotes cell survival and delays motor impairment in Niemann-Pick type A disease. Neurobiology of Disease, 2020, 144, 105046.	4.4	7
146	New paradigms for the treatment of lysosomal storage diseases: targeting the endocannabinoid system as a therapeutic strategy. Orphanet Journal of Rare Diseases, 2021, 16, 151.	2.7	7
147	Morbidity and mortality associated with Farber disease and prospects for therapy. Expert Opinion on Orphan Drugs, 2017, 5, 717-726.	0.8	6
148	Structural and functional analysis of the ASM p.Ala359Asp mutant that causes acid sphingomyelinase deficiency. Biochemical and Biophysical Research Communications, 2016, 479, 496-501.	2.1	5
149	Using wholeâ€exome sequencing to investigate the genetic bases of lysosomal storage diseases of unknown etiology. Human Mutation, 2017, 38, 1491-1499.	2.5	5
150	SiO₂ stimulates macrophage stress to induce the transformation of lung fibroblasts into myofibroblasts and its relationship with the sphingomyelin metabolic pathway. Journal of Applied Toxicology, 2021, 41, 1584-1597.	2.8	5
151	Fluorescence-Based Selection of Gene-Corrected Hematopoietic Stem and Progenitor Cells From Acid Sphingomyelinase-Deficient Mice: Implications for Niemann-Pick Disease Gene Therapy and the Development of Improved Stem Cell Gene Transfer Procedures. Blood, 1999, 93, 80-86.	1.4	5
152	The Genetics of Sphingolipid Hydrolases and Sphingolipid Storage Diseases. Handbook of Experimental Pharmacology, 2013, , 3-32.	1.8	4
153	Growth Plate Pathology in the Mucopolysaccharidosis Type VI Rat Modelâ€"An Experimental and Computational Approach. Diagnostics, 2020, 10, 360.	2.6	3
154	Signalling Effects Induced by Acid Ceramidase in Human Epithelial Or Leukemic Cell Lines. Cellular Physiology and Biochemistry, 2019, 52, 1092-1102.	1.6	3
155	Neural gene therapy for inherited diseases with mental retardation: Principles and prospects. Mental Retardation and Developmental Disabilities Research Reviews, 1995, 1, 39-48.	3.6	1
156	The Niemannâ€"Pick Diseases. , 2015, , 313-320.		1
157	A New Fluorescent Method to Detect Sulfamidase Activity in Blood, Tissue Extracts and Dried Blood Spots. Journal of Inborn Errors of Metabolism and Screening, 0, 9, .	0.3	1
158	A63: Treatment of Arthritis in Animal Models of the Mucopolysaccharidoses Using a Novel Anti-Inflammatory Drug, Pentosan Polysulfate. Arthritis and Rheumatology, 2014, 66, S93-S93.	5.6	0
159	Lysosomal Storage Diseases. , 2018, , 740-746.		0
160	The Niemannâ€"Pick diseases. , 2020, , 451-460.		0