Frances M Platt

List of Publications by Year
in descending order

205 papers	17,781 citations	13099 68 h-index	14759 127 g-index
222 all docs	222 docs citations	222 times ranked	18101 citing authors

#	Article	IF	Citations
1	Vesicle cholesterol controls exocytotic fusion pore. Cell Calcium, 2022, 101, 102503.	2.4	13
2	Glycosphingolipid metabolism and its role in ageing and Parkinsonâ€™s disease. Glycoconjugate Journal, 2022, 39, 39-53.	2.7	18
3	Correlation of age of onset and clinical severity in Niemannâ€“Pick disease type C1 with lysosomal abnormalities and gene expression. Scientific Reports, 2022, 12, 2162.	3.3	3
4	Current methods to analyze lysosome morphology, positioning, motility and function. Traffic, 2022, 23, 238-269.	2.7	37
5	Increasing Diversity in Admissions to Postgraduate Study. Journal of Medicinal Chemistry, 2022, 65, 5867-5869.	6.4	0
6	Glucosamine amends CNS pathology in mucopolysaccharidosis IIIC mouse expressing misfolded HGSNAT. Journal of Experimental Medicine, 2022, 219, .	8.5	7
7	Acetyl-leucine slows disease progression in lysosomal storage disorders. Brain Communications, 2021, 3, fcaa148.	3.3	37
8	Lipid-mediated motor-adaptor sequestration impairs axonal lysosome delivery leading to autophagic stress and dystrophy in Niemann-Pick type C. Developmental Cell, 2021, 56, 1452-1468.e8.	7.0	41
9	Lipid-mediated impairment of axonal lysosome transport contributing to autophagic stress. Autophagy, 2021, 17, 1796-1798.	9.1	10
10	Transcriptome of HPÎ²CD-treated Niemann-Pick disease type C1 cells highlights GPNMB as a biomarker for therapeutics. Human Molecular Genetics, 2021, 30, 2456-2468.	2.9	15
11	An iPSC model of hereditary sensory neuropathy-1 reveals L-serine-responsive deficits in neuronal ganglioside composition and axoglial interactions. Cell Reports Medicine, 2021, 2, 100345.	6.5	11
12	Acetylation turns leucine into a drug by membrane transporter switching. Scientific Reports, 2021, 11, 15812.	3.3	16
13	A modified density gradient proteomic-based method to analyze endolysosomal proteins in cardiac tissue. IScience, 2021, 24, 102949.	4.1	1
14	GM1 Gangliosidosisâ€”A Mini-Review. Frontiers in Genetics, 2021, 12, 734878.	2.3	38
15	Selective estrogen receptor modulators (SERMs) affect cholesterol homeostasis through the master regulators SREBP and LXR. Biomedicine and Pharmacotherapy, 2021, 141, 111871.	5.6	13
16	Identification of genetic modifiers of murine hepatic Î²-glucocerebrosidase activity. Biochemistry and Biophysics Reports, 2021, 28, 101105.	1.3	4
17	International consensus on clinical severity scale use in evaluating Niemannâ€“Pick disease Type C in paediatric and adult patients: results from a Delphi Study. Orphanet Journal of Rare Diseases, 2021, 16, 482.	2.7	8
18	Mechanistic convergence and shared therapeutic targets in Niemannâ€Pick disease. Journal of Inherited Metabolic Disease, 2020, 43, 574-585.	3.6	13

#	Article	IF	Citations
19	NPC1 Deficiency in Mice is Associated with Fetal Growth Restriction, Neonatal Lethality and Abnormal Lung Pathology. Journal of Clinical Medicine, 2020, 9, 12.	2.4	16
20	Systemic AAV9 gene therapy using the synapsin I promoter rescues a mouse model of neuronopathic Gaucher disease but with limited cross-correction potential to astrocytes. Human Molecular Genetics, 2020, 29, 1933-1949.	2.9	24
21	Sandhoff Disease: Improvement of Gait by Acetyl-DL-Leucine: A Case Report. Neuropediatrics, 2020, 51, 450-452.	0.6	11
22	Upregulating Î²-hexosaminidase activity in rodents prevents Î±-synuclein lipid associations and protects dopaminergic neurons from Î±-synuclein-mediated neurotoxicity. Acta Neuropathologica Communications, 2020, 8, 127.	5.2	17
23	Investigating the Mechanism of Cyclodextrins in the Treatment of Niemannâ€Pick Disease Type C Using Crosslinked 2â€Hydroxypropylâ€Î²â€cyclodextrin. Small, 2020, 16, e2004735.	10.0	16
24	c-Abl Inhibition Activates TFEB and Promotes Cellular Clearance in a Lysosomal Disorder. IScience, 2020, 23, 101691.	4.1	30
25	Molecular basis for a new bovine model of Niemann-Pick type C disease. PLoS ONE, 2020, 15, e0238697.	2.5	4
26	Defective platelet function in <scp>Niemannâ€Pick</scp> disease type <scp>C1</scp>. JIMD Reports, 2020, 56, 46-57.	1.5	9
27	Unexpected differences in the pharmacokinetics of N-acetyl-DL-leucine enantiomers after oral dosing and their clinical relevance. PLoS ONE, 2020, 15, e0229585.	2.5	21
28	Brain Pathology in Mucopolysaccharidoses (MPS) Patients with Neurological Forms. Journal of Clinical Medicine, 2020, 9, 396.	2.4	40
29	Beneficial Effects of Acetyl-DL-Leucine (ADLL) in a Mouse Model of Sandhoff Disease. Journal of Clinical Medicine, 2020, 9, 1050.	2.4	26
30	Metabolomic Studies of Lipid Storage Disorders, with Special Reference to Niemann-Pick Type C Disease: A Critical Review with Future Perspectives. International Journal of Molecular Sciences, 2020, 21, 2533.	4.1	13
31	Genetic background modifies phenotypic severity and longevity in a mouse model of Niemann-Pick disease type C1. DMM Disease Models and Mechanisms, 2020, 13, .	2.4	17
32	Unbiased yeast screens identify cellular pathways affected in Niemannâ€“Pick disease type C. Life Science Alliance, 2020, 3, e201800253.	2.8	10
33	Title is missing!. , 2020, 15, e0229585.		0
34	Title is missing!. , 2020, 15, e0229585.		0
35	Title is missing!. , 2020, 15, e0229585.		0
36	Title is missing!. , 2020, 15, e0229585.		0

#	Article	IF	Citations
37	Sterile activation of invariant natural killer T cells by ER-stressed antigen-presenting cells. Proceedings of the National Academy of Sciences of the United States of America, 2019, 116, 23671-23681.	7.1	21
38	Reduced sphingolipid hydrolase activities, substrate accumulation and ganglioside decline in Parkinsonâ€™s disease. Molecular Neurodegeneration, 2019, 14, 40.	10.8	100
39	Combined Anti-inflammatory and Neuroprotective Treatments Have the Potential to Impact Disease Phenotypes in Cln3âˆ’/âˆ’ Mice. Frontiers in Neurology, 2019, 10, 963.	2.4	13
40	Drugâ€induced increase in lysobisphosphatidic acid reduces the cholesterol overload in Niemannâ€“Pick type C cells and mice. EMBO Reports, 2019, 20, e47055.	4.5	33
41	Synthesis and Study of Multifunctional Cyclodextrinâ€“Deferasirox Hybrids. ChemMedChem, 2019, 14, 1484-1492.	3.2	5
42	TLR9-mediated dendritic cell activation uncovers mammalian ganglioside species with specific ceramide backbones that activate invariant natural killer T cells. PLoS Biology, 2019, 17, e3000169.	5.6	24
43	Age-related gait standards for healthy children and young people: the GOS-ICH paediatric gait centiles. Archives of Disease in Childhood, 2019, 104, 755-760.	1.9	10
44	Imaging of changes in copper trafficking and redistribution in a mouse model of Niemann-Pick C disease using positron emission tomography. BioMetals, 2019, 32, 293-306.	4.1	7
45	Glycosphingolipid levels and glucocerebrosidase activity are altered in normal aging of the mouse brain. Neurobiology of Aging, 2018, 67, 189-200.	3.1	66
46	Emptying the stores: lysosomal diseases and therapeutic strategies. Nature Reviews Drug Discovery, 2018, 17, 133-150.	46.4	191
47	Lysosomal storage diseases. Nature Reviews Disease Primers, 2018, 4, 27.	30.5	563
48	Altered Expression of Ganglioside Metabolizing Enzymes Results in GM3 Ganglioside Accumulation in Cerebellar Cells of a Mouse Model of Juvenile Neuronal Ceroid Lipofuscinosis. International Journal of Molecular Sciences, 2018, 19, 625.	4.1	12
49	Fetal gene therapy for neurodegenerative disease of infants. Nature Medicine, 2018, 24, 1317-1323.	30.7	117
50	Annual severity increment score as a tool for stratifying patients with Niemann-Pick disease type C and for recruitment to clinical trials. Orphanet Journal of Rare Diseases, 2018, 13, 143.	2.7	41
51	GM1 ganglioside-independent intoxication by Cholera toxin. PLoS Pathogens, 2018, 14, e1006862.	4.7	57
52	AAV9 intracerebroventricular gene therapy improves lifespan, locomotor function and pathology in a mouse model of Niemannâ€“Pick type C1 disease. Human Molecular Genetics, 2018, 27, 3079-3098.	2.9	51
53	A novel approach to analyze lysosomal dysfunctions through subcellular proteomics and lipidomics: the case of NPC1 deficiency. Scientific Reports, 2017, 7, 41408.	3.3	93
54	FTY720/fingolimod increases NPC1 and NPC2 expression and reduces cholesterol and sphingolipid accumulation in Niemannâ€Pick type C mutant fibroblasts. FASEB Journal, 2017, 31, 1719-1730.	0.5	39

#	Article	IF	Citations
55	Impaired antibacterial autophagy links granulomatous intestinal inflammation in Niemannâ€“Pick disease type C1 and XIAP deficiency with NOD2 variants in Crohn's disease. Gut, 2017, 66, 1060-1073.	12.1	126
56	Neuraminidases 3 and 4 regulate neuronal function by catabolizing brain gangliosides. FASEB Journal, 2017, 31, 3467-3483.	0.5	46
57	The metabolism of glucocerebrosides â€” From 1965 to the present. Molecular Genetics and Metabolism, 2017, 120, 22-26.	1.1	28
58	Inhibition of Î²-Glucocerebrosidase Activity Preserves Motor Unit Integrity in a Mouse Model of Amyotrophic Lateral Sclerosis. Scientific Reports, 2017, 7, 5235.	3.3	53
59	NMR analysis reveals significant differences in the plasma metabolic profiles of Niemann Pick C1 patients, heterozygous carriers, and healthy controls. Scientific Reports, 2017, 7, 6320.	3.3	17
60	Haematopoietic Stem Cell Transplantation Arrests theÂProgression of Neurodegenerative Disease in Late-Onset Tay-Sachs Disease. JIMD Reports, 2017, 41, 17-23.	1.5	18
61	N-Butyl-<scp>l</scp>-deoxynojirimycin (<scp>l</scp>-NBDNJ): Synthesis of an Allosteric Enhancer of Î±-Glucosidase Activity for the Treatment of Pompe Disease. Journal of Medicinal Chemistry, 2017, 60, 9462-9469.	6.4	31
62	Case Report: Ursodeoxycholic acid treatment in Niemann-Pick disease type C; clinical experience in four cases. Wellcome Open Research, 2017, 2, 75.	1.8	11
63	Differential response of the liver to bile acid treatment in a mouse model of Niemann-Pick disease type C. Wellcome Open Research, 2017, 2, 76.	1.8	2
64	Silencing the porcine iGb3s gene does not affect GalÎ±3Gal levels or measures of anticipated pigâ€toâ€human and pigâ€toâ€primate acute rejection. Xenotransplantation, 2016, 23, 106-116.	2.8	25
65	Identification of novel bile acids as biomarkers for the early diagnosis of Niemannâ€Pick C disease. FEBS Letters, 2016, 590, 1651-1662.	2.8	82
66	Chemoenzymatic Synthesis of a Phosphorylated Glycoprotein. Angewandte Chemie - International Edition, 2016, 55, 5058-5061.	13.8	46
67	Chemoenzymatic Synthesis of a Phosphorylated Glycoprotein. Angewandte Chemie, 2016, 128, 5142-5145.	2.0	8
68	An anecdotal report by an Oxford basic neuroscientist: effects of acetyl-dl-leucine on cognitive function and mobility in the elderly. Journal of Neurology, 2016, 263, 1239-1240.	3.6	7
69	Circadian profiling in two mouse models of lysosomal storage disorders; Niemann Pick type-C and Sandhoff disease. Behavioural Brain Research, 2016, 297, 213-223.	2.2	6
70	1H NMR-Linked Metabolomics Analysis of Liver from a Mouse Model of NP-C1 Disease. Journal of Proteome Research, 2016, 15, 3511-3527.	3.7	13
71	Immune dysfunction in Niemannâ€Pick disease type C. Journal of Neurochemistry, 2016, 136, 74-80.	3.9	55
72	Heat shock proteinâ€“based therapy as a potential candidate for treating the sphingolipidoses. Science Translational Medicine, 2016, 8, 355ra118.	12.4	137

#	Article	IF	Citations
73	High incidence of unrecognized visceral/neurological late-onset Niemann-Pick disease, type C1, predicted by analysis of massively parallel sequencing data sets. Genetics in Medicine, 2016, 18, 41-48.	2.4	171
74	Urinary excretion and metabolism of miglustat and valproate in patients with Niemannâ€“Pick type C1 disease: One- and two-dimensional solution-state 1 H NMR studies. Journal of Pharmaceutical and Biomedical Analysis, 2016, 117, 276-288.	2.8	4
75	A comparative study on fluorescent cholesterol analogs as versatile cellular reporters. Journal of Lipid Research, 2016, 57, 299-309.	4.2	78
76	Pathogenic mycobacteria achieve cellular persistence by inhibiting the Niemann-Pick Type C disease cellular pathway. Wellcome Open Research, 2016, 1, 18.	1.8	26
77	Defective Cytochrome P450-Catalysed Drug Metabolism in Niemann-Pick Type C Disease. PLoS ONE, 2016, 11, e0152007.	2.5	22
78	Expression of Ca ²⁺ â€permeable twoâ€pore channels rescues <scp>NAADP</scp> signalling in <scp>TPC</scp> â€deficient cells. EMBO Journal, 2015, 34, 1743-1758.	7.8	144
79	Preferential Coupling of the NAADP Pathway to Exocytosis in T-Cells. Messenger (Los Angeles, Calif:) Tj ETQq1 1 0.784314 rgBT /Over	0.3	8
80	A novel, highly sensitive and specific biomarker for Niemann-Pick type C1 disease. Orphanet Journal of Rare Diseases, 2015, 10, 78.	2.7	105
81	Intracellular sphingosine releases calcium from lysosomes. ELife, 2015, 4, .	6.0	115
82	Measuring relative lysosomal volume for monitoring lysosomal storage diseases. Methods in Cell Biology, 2015, 126, 331-347.	1.1	4
83	Guidelines for incorporating scientific knowledge and practice on rare diseases into higher education: neuronal ceroid lipofuscinoses as a model disorder. Biochimica Et Biophysica Acta - Molecular Basis of Disease, 2015, 1852, 2316-2323.	3.8	13
84	Amyotrophic lateral sclerosis and denervation alter sphingolipids and up-regulate glucosylceramide synthase. Human Molecular Genetics, 2015, 24, 7390-7405.	2.9	84
85	Biomarkers for disease progression and AAV therapeutic efficacy in feline Sandhoff disease. Experimental Neurology, 2015, 263, 102-112.	4.1	26
86	Bridging the age spectrum of neurodegenerative storage diseases. Best Practice and Research in Clinical Endocrinology and Metabolism, 2015, 29, 127-143.	4.7	18
87	A Novel Mouse Model of a Patient Mucolipidosis II Mutation Recapitulates Disease Pathology. Journal of Biological Chemistry, 2014, 289, 26709-26721.	3.4	16
88	Effects of miglustat treatment in a patient affected by an atypical form of Tangier disease. Orphanet Journal of Rare Diseases, 2014, 9, 143.	2.7	14
89	Improved neuroprotection using miglustat, curcumin and ibuprofen as a triple combination therapy in Niemannâ€“Pick disease type C1 mice. Neurobiology of Disease, 2014, 67, 9-17.	4.4	74
90	Hepatic metabolic response to restricted copper intake in a Niemannâ€“Pick C murine model. Metallomics, 2014, 6, 1527-1539.	2.4	11

#	Article	IF	Citations
91	Disorders of Cholesterol Metabolism and Their Unanticipated Convergent Mechanisms of Disease. Annual Review of Genomics and Human Genetics, 2014, 15, 173-194.	6.2	73
92	RIPK3 as a potential therapeutic target for Gaucher's disease. Nature Medicine, 2014, 20, 204-208.	30.7	147
93	Sphingolipid lysosomal storage disorders. Nature, 2014, 510, 68-75.	27.8	270
94	Altered distribution and function of natural killer cells in murine and human Niemann-Pick disease type C1. Blood, 2014, 123, 51-60.	1.4	38
95	Relative acidic compartment volume as a lysosomal storage disorderâ€“associated biomarker. Journal of Clinical Investigation, 2014, 124, 1320-1328.	8.2	63
96	¹H NMR-Linked Urinary Metabolic Profiling of Niemann-Pick Class C1 (NPC1) Disease: Identification of Potential New Biomarkers using Correlated Component Regression (CCR) and Genetic Algorithm (GA) Analysis Strategies. Current Metabolomics, 2014, 2, 88-121.	0.5	14
97	Glycomimetic affinity-enrichment proteomics identifies partners for a clinically-utilized iminosugar. Chemical Science, 2013, 4, 3442.	7.4	7
98	Vesicular and non-vesicular transport feed distinct glycosylation pathways in the Golgi. Nature, 2013, 501, 116-120.	27.8	136
99	Î²-Glucosidase 2 (GBA2) Activity and Imino Sugar Pharmacology. Journal of Biological Chemistry, 2013, 288, 26052-26066.	3.4	69
100	Cyclodextrin alleviates neuronal storage of cholesterol in Niemannâ€Pick C disease without evidence of detectable bloodâ€“brain barrier permeability. Journal of Inherited Metabolic Disease, 2013, 36, 491-498.	3.6	74
101	Mutations in B4GALNT1 (GM2 synthase) underlie a new disorder of ganglioside biosynthesis. Brain, 2013, 136, 3618-3624.	7.6	115
102	The Yeast P5 Type ATPase, Spf1, Regulates Manganese Transport into the Endoplasmic Reticulum. PLoS ONE, 2013, 8, e85519.	2.5	62
103	Globosides but Not Isoglobosides Can Impact the Development of Invariant NKT Cells and Their Interaction with Dendritic Cells. Journal of Immunology, 2012, 189, 3007-3017.	0.8	38
104	Lysosomal storage disorders: The cellular impact of lysosomal dysfunction. Journal of Cell Biology, 2012, 199, 723-734.	5.2	579
105	Invariant natural killer <scp>T</scp> cells are not affected by lysosomal storage in patients with <scp>N</scp>iemannâ€<scp>P</scp>ick disease type <scp>C</scp>. European Journal of Immunology, 2012, 42, 1886-1892.	2.9	14
106	Early glial activation, synaptic changes and axonal pathology in the thalamocortical system of Niemannâ€“Pick type C1 mice. Neurobiology of Disease, 2012, 45, 1086-1100.	4.4	84
107	Molecular mechanisms of endolysosomal Ca2+ signalling in health and disease. Biochemical Journal, 2011, 439, 349-378.	3.7	329
108	Lysosomal Ca2+ homeostasis: Role in pathogenesis of lysosomal storage diseases. Cell Calcium, 2011, 50, 200-205.	2.4	122

#	Article	IF	Citations
109	A sensitive and specific LC-MS/MS method for rapid diagnosis of Niemann-Pick C1 disease from human plasma. Journal of Lipid Research, 2011, 52, 1435-1445.	4.2	230
110	Diverse Endogenous Antigens for Mouse NKT Cells: Self-Antigens That Are Not Glycosphingolipids. Journal of Immunology, 2011, 186, 1348-1360.	0.8	54
111	Glycosphingolipid storage leads to the enhanced degradation of the B cell receptor in Sandhoff disease mice. Journal of Inherited Metabolic Disease, 2010, 33, 261-270.	3.6	12
112	Purified TPC Isoforms Form NAADP Receptors with Distinct Roles for Ca2+ Signaling and Endolysosomal Trafficking. Current Biology, 2010, 20, 703-709.	3.9	234
113	Restricted ketogenic diet enhances the therapeutic action of Nâ€butyldeoxynojirimycin towards brain GM2 accumulation in adult Sandhoff disease mice. Journal of Neurochemistry, 2010, 113, 1525-1535.	3.9	23
114	Lipids on Trial: The Search for the Offending Metabolite in Niemann-Pick type C Disease. Traffic, 2010, 11, 419-428.	2.7	170
115	Macroautophagy Is Not Directly Involved in the Metabolism of Amyloid Precursor Protein. Journal of Biological Chemistry, 2010, 285, 37415-37426.	3.4	81
116	Common and Uncommon Pathogenic Cascades in Lysosomal Storage Diseases. Journal of Biological Chemistry, 2010, 285, 20423-20427.	3.4	298
117	Endolysosomal calcium regulation and disease. Biochemical Society Transactions, 2010, 38, 1458-1464.	3.4	56
118	Critical role of iron in the pathogenesis of the murine gangliosidoses. Neurobiology of Disease, 2009, 34, 406-416.	4.4	29
119	Beneficial effects of anti-inflammatory therapy in a mouse model of Niemann-Pick disease type C1. Neurobiology of Disease, 2009, 36, 242-251.	4.4	132
120	Treating lysosomal storage disorders: Current practice and future prospects. Biochimica Et Biophysica Acta - Molecular Cell Research, 2009, 1793, 737-745.	4.1	76
121	Neural Stem Cell Transplantation Benefits a Monogenic Neurometabolic Disorder During the Symptomatic Phase of Disease. Stem Cells, 2009, 27, 2362-2370.	3.2	44
122	A new surrogate marker for CNS pathology in Niemannâ€“Pick disease type C?. Molecular Genetics and Metabolism, 2009, 96, 53-54.	1.1	3
123	CD1d presentation of glycolipids. Immunology and Cell Biology, 2008, 86, 588-597.	2.3	21
124	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
125	Substrate reduction therapy. Acta Paediatrica, International Journal of Paediatrics, 2008, 97, 88-93.	1.5	139
126	Glycosphingolipid depletion in PC12 cells using iminosugars protects neuronal membranes from anti-ganglioside antibody mediated injury. Journal of Neuroimmunology, 2008, 203, 33-38.	2.3	3

#	Article	IF	Citations
127	Autophagy Induction and Autophagosome Clearance in Neurons: Relationship to Autophagic Pathology in Alzheimer's Disease. Journal of Neuroscience, 2008, 28, 6926-6937.	3.6	979
128	N-butyldeoxygalactonojirimycin reduces brain ganglioside and GM2 content in neonatal Sandhoff disease mice. Neurochemistry International, 2008, 52, 1125-1133.	3.8	44
129	Beneficial effects of substrate reduction therapy in a mouse model of GM1 gangliosidosis. Molecular Genetics and Metabolism, 2008, 94, 204-211.	1.1	75
130	Male Germ Cells Require Polyenoic Sphingolipids with Complex Glycosylation for Completion of Meiosis. Journal of Biological Chemistry, 2008, 283, 13357-13369.	3.4	100
131	Differential sensitivity of mouse strains to an N-alkylated imino sugar: glycosphingolipid metabolism and acrosome formation. Pharmacogenomics, 2008, 9, 717-731.	1.3	8
132	Invariant NKT cells reduce the immunosuppressive activity of influenza A virusâ€“induced myeloid-derived suppressor cells in mice and humans. Journal of Clinical Investigation, 2008, 118, 4036-4048.	8.2	299
133	Normal development and function of invariant natural killer T cells in mice with isoglobotrihexosylceramide (iGb3) deficiency. Proceedings of the National Academy of Sciences of the United States of America, 2007, 104, 5977-5982.	7.1	198
134	Accumulation of Glucosylceramide in Murine Testis, Caused by Inhibition of Î²-Glucosidase 2. Journal of Biological Chemistry, 2007, 282, 32655-32664.	3.4	63
135	Modulation of human natural killer T cell ligands on TLR-mediated antigen-presenting cell activation. Proceedings of the National Academy of Sciences of the United States of America, 2007, 104, 20490-20495.	7.1	173
136	Implications for invariant natural killer T cell ligands due to the restricted presence of isoglobotrihexosylceramide in mammals. Proceedings of the National Academy of Sciences of the United States of America, 2007, 104, 5971-5976.	7.1	145
137	The postacrosomal assembly of sperm head protein, PAWP, is independent of acrosome formation and dependent on microtubular manchette transport. Developmental Biology, 2007, 312, 471-483.	2.0	64
138	Activation of Invariant NKT Cells by Toll-like Receptor 9-Stimulated Dendritic Cells Requires Type I Interferon and Charged Glycosphingolipids. Immunity, 2007, 27, 597-609.	14.3	243
139	The sensitivity of murine spermiogenesis to miglustat is a quantitative trait: a pharmacogenetic study. Reproductive Biology and Endocrinology, 2007, 5, 1.	3.3	35
140	Stem cells act through multiple mechanisms to benefit mice with neurodegenerative metabolic disease. Nature Medicine, 2007, 13, 439-447.	30.7	293
141	Glycosphingolipid synthesis requires FAPP2 transfer of glucosylceramide. Nature, 2007, 449, 62-67.	27.8	359
142	Substrate Reduction Therapy. , 2007, , 153-168.		5
143	Activation of Invariant NKT Cells by the Helminth Parasite Schistosoma mansoni. Journal of Immunology, 2006, 176, 2476-2485.	0.8	78
144	Glycolipid receptor depletion as an approach to specific antimicrobial therapy. FEMS Microbiology Letters, 2006, 258, 1-8.	1.8	18

#	Article	IF	Citations
145	The Association of Shiga-like Toxin with Detergent-resistant Membranes Is Modulated by Glucosylceramide and Is an Essential Requirement in the Endoplasmic Reticulum for a Cytotoxic Effect. Molecular Biology of the Cell, 2006, 17, 1375-1387.	2.1	93
146	Long-term non-hormonal male contraception in mice using N-butyldeoxynojirimycin. Human Reproduction, 2006, 21, 1309-1315.	0.9	25
147	Impaired selection of invariant natural killer T cells in diverse mouse models of glycosphingolipid lysosomal storage diseases. Journal of Experimental Medicine, 2006, 203, 2293-2303.	8.5	127
148	Storage solutions: treating lysosomal disorders of the brain. Nature Reviews Neuroscience, 2005, 6, 713-725.	10.2	159
149	Imino sugar inhibitors for treating the lysosomal glycosphingolipidoses. Glycobiology, 2005, 15, 43R-52R.	2.5	204
150	Alkylated Imino Sugars, Reversible Male Infertility-Inducing Agents, Do Not Affect the Genetic Integrity of Male Mouse Germ Cells During Short-Term Treatment Despite Induction of Sperm Deformities1. Biology of Reproduction, 2005, 72, 805-813.	2.7	55
151	New Developments in Treating Glycosphingolipid Storage Diseases. Advances in Experimental Medicine and Biology, 2005, 564, 117-126.	1.6	12
152	Inhibition of Glucosylceramide Synthase Does Not Reverse Drug Resistance in Cancer Cells. Journal of Biological Chemistry, 2004, 279, 40412-40418.	3.4	48
153	Accumulation of Glycosphingolipids in Niemann-Pick C Disease Disrupts Endosomal Transport. Journal of Biological Chemistry, 2004, 279, 26167-26175.	3.4	167
154	Inhibition of Î±-Glucosidases I and II Increases the Cell Surface Expression of Functional Class A Macrophage Scavenger Receptor (SR-A) by Extending Its Half-life. Journal of Biological Chemistry, 2004, 279, 39303-39309.	3.4	15
155	Infantile-onset symptomatic epilepsy syndrome caused by a homozygous loss-of-function mutation of GM3 synthase. Nature Genetics, 2004, 36, 1225-1229.	21.4	359
156	Inhibition of glycogen breakdown by imino sugars in vitro and in vivo. Biochemical Pharmacology, 2004, 67, 697-705.	4.4	23
157	Analysis of fluorescently labeled glycosphingolipid-derived oligosaccharides following ceramide glycanase digestion and anthranilic acid labeling. Analytical Biochemistry, 2004, 331, 275-282.	2.4	162
158	N-butyldeoxygalactonojirimycin reduces neonatal brain ganglioside content in a mouse model of GM1 gangliosidosis. Journal of Neurochemistry, 2004, 89, 645-653.	3.9	68
159	NSAIDs increase survival in the Sandhoff disease mouse: Synergy withN-butyldeoxynojirimycin. Annals of Neurology, 2004, 56, 642-649.	5.3	116
160	Miglustat: profile report. Drugs and Therapy Perspectives, 2004, 20, 5-7.	0.6	0
161	Therapy of Niemannâ€“Pick disease, type C. Biochimica Et Biophysica Acta - Molecular and Cell Biology of Lipids, 2004, 1685, 77-82.	2.4	60
162	Glycosphingolipids in endocytic membrane transport. Seminars in Cell and Developmental Biology, 2004, 15, 409-416.	5.0	28

#	Article	IF	Citations
163	Improved outcome of N-butyldeoxygalactonojirimycin-mediated substrate reduction therapy in a mouse model of Sandhoff disease. Neurobiology of Disease, 2004, 16, 506-515.	4.4	79
164	Treatment with miglustat reverses the lipid-trafficking defect in Niemannâ€“Pick disease type C. Neurobiology of Disease, 2004, 16, 654-658.	4.4	147
165	Cellular effects of deoxynojirimycin analogues: inhibition of N-linked oligosaccharide processing and generation of free glucosylated oligosaccharides. Biochemical Journal, 2004, 381, 867-875.	3.7	39
166	Cellular effects of deoxynojirimycin analogues: uptake, retention and inhibition of glycosphingolipid biosynthesis. Biochemical Journal, 2004, 381, 861-866.	3.7	45
167	Lysosomal defects and storage. , 2004, , 32-49.		23
168	Inhibition of substrate synthesis: a pharmacological approach for glycosphingolipid storage disease therapy. , 2004, , 381-408.		11
169	Storage diseases: new insights into sphingolipid functions. Trends in Cell Biology, 2003, 13, 195-203.	7.9	32
170	Glycolipid depletion in antimicrobial therapy. Molecular Microbiology, 2003, 47, 453-461.	2.5	39
171	Miglustat. Drugs, 2003, 63, 2435-2436.	10.9	1
172	Smallâ€“molecule therapeutics for the treatment of glycolipid lysosomal storage disorders. Philosophical Transactions of the Royal Society B: Biological Sciences, 2003, 358, 927-945.	4.0	58
173	Substrate reduction therapy in mouse models of the glycosphingolipidoses. Philosophical Transactions of the Royal Society B: Biological Sciences, 2003, 358, 947-954.	4.0	50
174	Inhibition of Calcium Uptake via the Sarco/Endoplasmic Reticulum Ca2+-ATPase in a Mouse Model of Sandhoff Disease and Prevention by Treatment with N-Butyldeoxynojirimycin. Journal of Biological Chemistry, 2003, 278, 29496-29501.	3.4	129
175	Therapeutic Applications of Imino Sugars in Lysosomal Storage Disorders. Current Topics in Medicinal Chemistry, 2003, 3, 561-574.	2.1	156
176	Membrane disruption and cytotoxicity of hydrophobic N-alkylated imino sugars is independent of the inhibition of protein and lipid glycosylation. Biochemical Journal, 2003, 374, 307-314.	3.7	54
177	Reversible infertility in male mice after oral administration of alkylated imino sugars: A nonhormonal approach to male contraception. Proceedings of the National Academy of Sciences of the United States of America, 2002, 99, 17173-17178.	7.1	104
178	Glucosylceramide modulates membrane traffic along the endocytic pathway. Journal of Lipid Research, 2002, 43, 1837-1845.	4.2	120
179	Preparation, biochemical characterization and biological properties of radiolabelled N-alkylated deoxynojirimycins. Biochemical Journal, 2002, 366, 225-233.	3.7	75
180	An Inducible Mouse Model of Late Onset Tayâ€“Sachs Disease. Neurobiology of Disease, 2002, 10, 201-210.	4.4	31

#	Article	IF	Citations
181	Modulation of THP-1 Macrophage and Cholesterol-Loaded Foam Cell Apolipoprotein E Levels by Glycosphingolipids. Biochemical and Biophysical Research Communications, 2002, 290, 1361-1367.	2.1	30
182	Inhibition of Glycosphingolipid Biosynthesis Does Not Impair Growth or Morphogenesis of the Postimplantation Mouse Embryo. Journal of Neurochemistry, 2002, 70, 871-882.	3.9	28
183	Targeting glycosylation as a therapeutic approach. Nature Reviews Drug Discovery, 2002, 1, 65-75.	46.4	409
184	Increased glycosphingolipid levels in serum and aortae of apolipoprotein E gene knockout mice. Journal of Lipid Research, 2002, 43, 205-214.	4.2	56
185	Increased glycosphingolipid levels in serum and aortae of apolipoprotein E gene knockout mice. Journal of Lipid Research, 2002, 43, 205-14.	4.2	50
186	Enhanced survival in Sandhoff disease mice receiving a combination of substrate deprivation therapy and bone marrow transplantation. Blood, 2001, 97, 327-329.	1.4	118
187	Carbohydrate Receptor Depletion as an Antimicrobial Strategy for Prevention of Urinary Tract Infection. Journal of Infectious Diseases, 2001, 183, S70-S73.	4.0	13
188	Substrate reduction therapy for glycosphingolipid storage disorders. Expert Opinion on Investigational Drugs, 2001, 10, 455-466.	4.1	72
189	N-butyldeoxygalactonojirimycin: a more selective inhibitor of glycosphingolipid biosynthesis than N-butyldeoxynojirimycin, in vitro and in vivo. Biochemical Pharmacology, 2000, 59, 821-829.	4.4	137
190	Substrate deprivation: A new therapeutic approach for the glycosphingolipid lysosomal storage diseases. Expert Reviews in Molecular Medicine, 2000, 2, 1-17.	3.9	12
191	Novel oral treatment of Gaucher's disease with N-butyldeoxynojirimycin (OGT 918) to decrease substrate biosynthesis. Lancet, The, 2000, 355, 1481-1485.	13.7	700
192	Inhibition of Glycosphingolipid Biosynthesis:Â Application to Lysosomal Storage Disorders. Chemical Reviews, 2000, 100, 4683-4696.	47.7	290
193	Inhibition of N-Glycan Processing in B16 Melanoma Cells Results in Inactivation of Tyrosinase but Does Not Prevent Its Transport to the Melanosome. Journal of Biological Chemistry, 1997, 272, 15796-15803.	3.4	76
194	Extensive Glycosphingolipid Depletion in the Liver and Lymphoid Organs of Mice Treated with N-Butyldeoxynojirimycin. Journal of Biological Chemistry, 1997, 272, 19365-19372.	3.4	115
195	Ultrastructural changes in the Golgi apparatus and secretory granules of HL-60 cells treated with the imino sugar N-butyldeoxynojirimycin. Biology of the Cell, 1997, 89, 123-131.	2.0	3
196	Ultrastructural changes in the Golgi apparatus and secretory granules of HL-60 cells treated with the imino sugar N-butyldeoxynojirimycin. Biology of the Cell, 1997, 89, 123-131.	2.0	1
197	Inhibitors of Glycosphingolipid Biosynthesis.. Trends in Glycoscience and Glycotechnology, 1995, 7, 495-511.	0.1	38
198	Monoclonal antibodies specific for novel murine cell surface markers define subpopulations of germinal center cells. Cellular Immunology, 1992, 143, 449-466.	3.0	7

#	Article	IF	Citations
199	Modulation of cell-surface transferrin receptor by the imino sugar N-butyldeoxynojirimycin. FEBS Journal, 1992, 208, 187-193.	0.2	20
200	Analysis and isolation of human transferrin receptor using the OKT-9 monoclonal antibody covalently crosslinked to magnetic beads. Analytical Biochemistry, 1991, 199, 219-222.	2.4	33
201	bcl-2-Immunoglobulin transgenic mice demonstrate extended B cell survival and follicular lymphoproliferation. Cell, 1989, 57, 79-88.	28.9	1,181
202	Medicinal use of Iminosugars. , 0, , 295-326.		9
203	Pathogenic mycobacteria achieve cellular persistence by inhibiting the Niemann-Pick Type C disease cellular pathway. Wellcome Open Research, 0, 1, 18.	1.8	30
204	Differential response of the liver to bile acid treatment in a mouse model of Niemann-Pick disease type C. Wellcome Open Research, 0, 2, 76.	1.8	1
205	Anti-TNF therapy for inflammatory bowel disease in patients with neurodegenerative Niemann-Pick disease Type C. Wellcome Open Research, 0, 7, 11.	1.8	3