

Sphingolipidoses

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Sphingolipidoses are a class of disorders relating to Sphingolipid metabolism. Examples include: * Fabry's disease * Gangliosidosis ** GM1 gangliosidosis ** GM2 gangliosidosis *** Tay-Sachs disease *** Sandhoff disease * Gaucher's disease * Krabbe disease * Metachromatic leukodystrophy * Niemann-Pick disease ([From the Wikipedia article Sphingolipidoses](#).)

Recent Publications on Sphingolipidoses:



- [Musculoskeletal manifestations of lysosomal storage disorders.](#)
Lysosomal storage disorders (LSDs), a heterogeneous group of inborn...18th November, 2009
Department of Paediatric Immunology/Haematology and SCT, University- Ann Rheum Dis. 2009 Nov;68(11):1659-65. ([DOI Direct Link](#))
- [Glycosphingolipids--nature, function, and pharmacological modulation.](#)
The discovery of the glycosphingolipids is generally attributed to Johan...29th October, 2009
Leiden Institute of Chemistry, Gorlaeus Laboratories, Leiden University,- Angew Chem Int Ed Engl. 2009;48(47):8848-69. ([DOI Direct Link](#))
- [Molecular facets of sphingolipids: mediators of diseases.](#)
Sphingolipids constitute a biologically active lipid class that is...22nd October, 2009
Department of Chemical Engineering, Bogazici University, Bebek, Istanbul,- Biotechnol J. 2009 Jul;4(7):1028-41. ([DOI Direct Link](#))
- [Cholesterol regulation of rab-mediated sphingolipid endocytosis.](#)
Despite a tight regulation of its intracellular content, cholesterol is...2nd October, 2009
VCAD System Research Program, Bio-research Infrastructure Construction- Glycoconj J. 2009 Aug;26(6):705-10. Epub 2008 Oct 8. ([DOI Direct Link](#))
- [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.](#)
Many lipidomic approaches focus on investigating aspects of sphingolipid...4th August, 2009
LIMES - Life and Medical Sciences Institute, Membrane Biology and Lipid- J Chromatogr B Analyt Technol Biomed Life Sci. 2009 Oct 1;877(27):2976-82. ([DOI Direct Link](#))
- [Transplantation of oligodendrocyte progenitor cells in animal models of leukodystrophies.](#)
Leukodystrophies represent a wide variety of hereditary disorders of the...30th June, 2009
Department of Medical Sciences, School of Veterinary Medicine, University- Methods Mol Biol. 2009;549:175-85. ([DOI Direct Link](#))
- [Secondary lipid accumulation in lysosomal disease.](#)
Lysosomal diseases are inherited metabolic disorders caused by defects in...6th June, 2009
Dominick P. Purpura Department of Neuroscience, Rose F. Kennedy Center,- Biochim Biophys Acta.

- 2009 Apr;1793(4):726-36. Epub 2008 Dec 9. ([DOI Direct Link](#))
- [Principles of lysosomal membrane degradation: Cellular topology and biochemistry of lysosomal lipid degradation.](#)
*Cellular membranes enter the lysosomal compartment by endocytosis...*6th June, 2009
 LIMES Program Unit Membrane Biology and Lipid Biochemistry, Laboratory of- Biochim Biophys Acta. 2009 Apr;1793(4):674-83. Epub 2008 Nov 1. ([DOI Direct Link](#))
 - [Prenatal diagnosis of perplexing cases of lipidoses.](#)
*OBJECTIVE: To present and discuss the technical, ethical and counseling...*6th March, 2009
 Gynecology and Obstetrics Department, Faculty of Medicine, Cairo- Bratisl Lek Listy. 2008;109(11):493-6.
 - [Megalencephalic leukodystrophy with simple hemihyperplasia: a rare association.](#)
*Lysosomal diseases are inherited metabolic disorders caused by defects in...*25th February, 2009
 Genetic unit, Department of Pediatrics, All India Institute of Medical- Clin Dysmorphol. 2009 Jan;18(1):49-51. ([DOI Direct Link](#))
 - [Regulation of lipid metabolism by sphingolipids.](#)
*Sphingolipids, together with phospholipids and cholesterol are key...*25th December, 2008
 Department of Pathology, Columbia University, 168 W 168 St, BB 457, New- Subcell Biochem. 2008;49:371-85. ([DOI Direct Link](#))
 - [Arsenic induces oxidative stress, sphingolipidosis, depletes proteins and some antioxidants in various regions of rat brain.](#)
*OBJECTIVES: To seek an interrelationship, if any, between oxidant stress...*17th December, 2008
 Department of Biochemistry, Nepalgunj Medical College, Nepalgunj, Nepal.- Kathmandu Univ Med J (KUMJ). 2008 Jan-Mar;6(1):60-9.
 - [Plasmalogen levels in Gaucher disease.](#)
*Plasmalogens represent a unique type of phospholipids characterized by the...*18th November, 2008
 Department of Enzymology and Cellular Function, Institute of Child Health,- Blood Cells Mol Dis. 2008 Sep-Oct;41(2):196-9. Epub 2008 May 23. ([DOI Direct Link](#))
 - [The sphingolipid salvage pathway in ceramide metabolism and signaling.](#)
*Sphingolipids are important components of eukaryotic cells, many of which...*30th September, 2008
 Department of Biochemistry and Molecular Biology, Medical University of- Cell Signal. 2008 Jun;20(6):1010-8. Epub 2007 Dec 14. ([DOI Direct Link](#))
 - [The C-terminal region of human adipose triglyceride lipase affects enzyme activity and lipid droplet binding.](#)
*Adipose triglyceride lipase (ATGL) catalyzes the first step in the...*26th July, 2008
 Institute of Molecular Biosciences, University of Graz, 8010 Graz,- J Biol Chem. 2008 Jun 20;283(25):17211-20. Epub 2008 Apr 29. ([DOI Direct Link](#))

Sphingolipidoses Patents:



- 4639437- [Kit or device and method for administering gangliosides and derivatives thereof by inhalation and pharmaceutical compositions suitable therefor](#)
- 5710028- [Method of quick screening and identification of specific DNA sequences by single nucleotide primer extension and kits therefor](#)
- 5714478- [Sphingosylphosphorylcholine as a wound-healing agent](#)

- 5773278- [Acid sphingomyelinase gene](#)
- 5792858- [Lysosphingolipid derivatives](#)
- 5827830- [Plasmalopsychosines and plasmalocerebrosides](#)
- 5866690- [Detection of malignant tumor cells](#)
- 5885786- [Methods for screening of substances for inhibition of multidrug resistance](#)
- 6090565- [Sphingoglycolipids as markers for multidrug resistant cancers](#)
- 6103694- [Method of decreasing radiation of radio-mimetic chemotherapy for hematopoietic pluripotent cell engraftment](#)
- 6177447- [Deoxynojirimycin derivatives and their uses as glucosylceramidase inhibitors](#)
- 5686240- [Acid sphingomyelinase gene and diagnosis of Niemann-Pick disease](#)
- 5610063- [cDNA for .alpha.-N-acetyl-galactosaminidase from Gallus domesticus](#)
- 5554608- [Inhibition of hair growth](#)
- 4716223- [Method for preparing ganglioside derivatives and use thereof in pharmaceutical compositions](#)
- 4816450- [Inhibition of protein kinase C by long-chain bases](#)
- 4866042- [Method for the delivery of genetic material across the blood brain barrier](#)
- 4937232- [Inhibition of protein kinase C by long-chain bases](#)
- 5137919- [Effect of N,N,N,-trimethylsphingosine on protein kinase C activity melanoma cell growth in vitro; metastatic potential in vivo and human platelet aggregation](#)
- 5248824- [Method of preparing N,N,N-trimethylsphingosine](#)
- 5512471- [Process for purifying an .alpha.-D-galactosidase isozyme from Coffea beans](#)
- 5519007- [Lysosphingolipid derivatives](#)
- 5532141- [Process for obtaining ganglioside lipids](#)
- 5549892- [Enhanced in vivo uptake of glucocerebrosidase](#)
- 6387366- [Methods for reducing adverse side effects associated with cellular transplantation](#)
- 6423311- [Method of decreasing radiation or radio-mimetic chemotherapy for hematopoietic pluripotent cell engraftment](#)
- 7427494- [Nucleic acid encoding cytokine receptor zcytor17](#)
- 7429460- [Methods of screening for inhibitors of phospholipid synthesis related to glycolipid-storage diseases](#)
- 7459293- [Cytokine zcytor17 ligand polynucleotides](#)
- 7485435- [Simultaneous detection of metabolic enzyme activity and metabolite levels](#)
- 7494804- [Polynucleotide encoding cytokine receptor zcytor17 multimer](#)
- 7495080- [Cytokine receptor zcytor17](#)
- 7507795- [Murine ZCYTOR17 ligand polypeptides](#)
- 7521537- [Cytokine receptor zcytor17](#)
- 7528153- [Deoxynojirimycin analogues and their uses as glucosylceramidase inhibitors](#)
- 7531636- [Antibodies to murine zcytor17 ligand](#)
- 7425325- [Cytokine zcytor17 ligand polypeptides](#)
- 7338781- [Organic anion transporting \(oat\)-like protein UST3-LIKE1 and uses thereof](#)
- 7329638- [Drug delivery compositions](#)
- 6541218- [Acid sphingomyelinase protein and methods of treating type B Niemann-Pick disease](#)
- 6693077- [Keratinocyte growth factor-2](#)
- 6846968- [Production of lysosomal enzymes in plants by transient expression](#)
- 6863885- [Method of decreasing radiation or radio-mimetic chemotherapy for hematopoietic pluripotent cell engraftment](#)
- 6887696- [Production of lysosomal enzymes in plants by transient expression](#)
- 6890748- [Production of lysosomal enzymes in plants by transient expression](#)
- 7064186- [Cytokine zcytor17 ligand](#)
- 7111346- [Reciprocating movement platform for the external addition of pulses of the fluid channels of a subject](#)
- 7228576- [Reciprocating movement platform for the external addition of pulses to the fluid channels of a subject](#)
- 7232667- [Keratinocyte growth factor-2 polynucleotides](#)

- 7563591- [Method for assaying the activity of lysosomal enzymes](#)

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