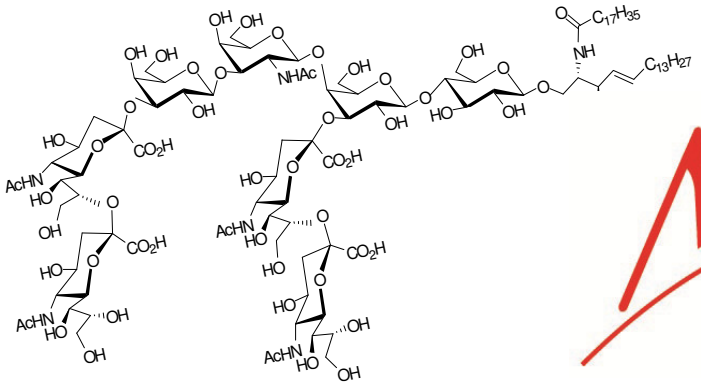


MATREYA LLC

Lipids and Biochemicals

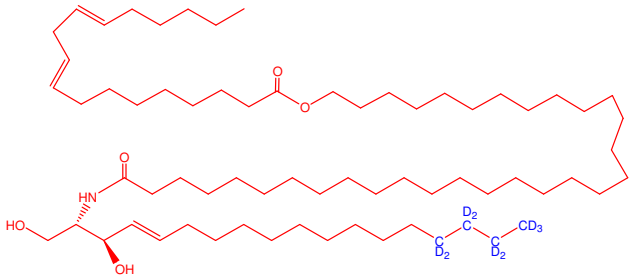
NEWSLETTER FOR GLYCO/SPHINGOLIPID RESEARCH

JANUARY 2019

Interactions of Ceramides and Proteins

Ceramides, ubiquitous sphingolipid intermediates with multiple physiological functions, often exert their powerful effects by direct interactions with proteins. Identified ceramide-binding proteins include the ceramide transfer protein CERT, the kinase suppressor of Ras, protein kinase c-Raf, cathepsin D, protein phosphatase 2A inhibitor SET, and StarD7.[1] By manipulating specific cardiac ceramide-binding proteins, researchers were able to reverse ceramide-induced lipotoxic cardiomyopathy.[2]

In addition to ceramides forming lose complexes with proteins, ceramides have been found to be attached, through ester linkages, to various proteins of the cornified cell envelope of the skin. During protein envelope assembly, the membrane is converted to a layer of ceramides covalently bound to the protein envelope on the extracellular surface. It is thought that the ester-linked ceramides constitute a hydrophobic surface that functions as a water barrier layer.[3]



N-(32-Linoleoyloxy-dotriacontanoyl)-sphingosine-D9
Cat.# 2208

References:

1. S. Bockelmann et al. A search for ceramide binding proteins using bifunctional lipid analogs yields CERT-related protein StarD7. J Lipid Res 59(3), 530 (2018)

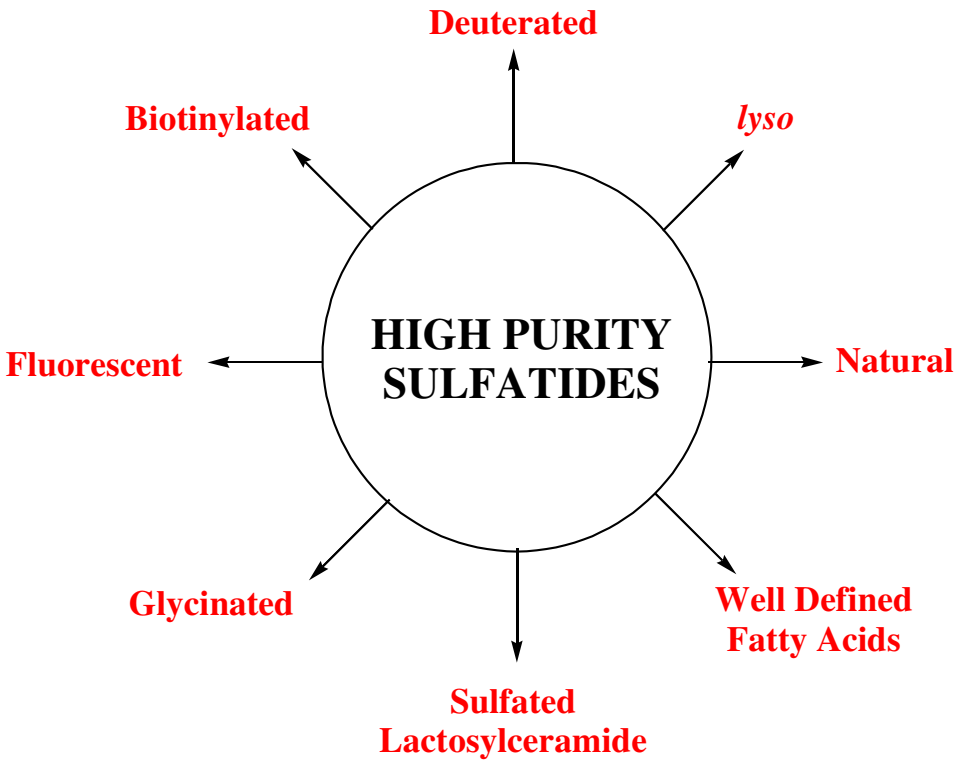
2. S. Walls et al. Ceramide-Protein Interactions Modulate Ceramide-Associated Lipotoxic Cardiomyopathy. Cell Rep. 22(10), 2702-2715 (2018)

3. L. Marekov and P. Steinert. Ceramides are bound to structural proteins of the human foreskin epidermal cornified cell envelope. J Biol Chem. 273(28), 17763-17770 (1998)

Please visit www.matreya.com for a full list of high purity ceramide standards

Inside This Issue

Sulfatides in Metachromatic Leukodystrophy



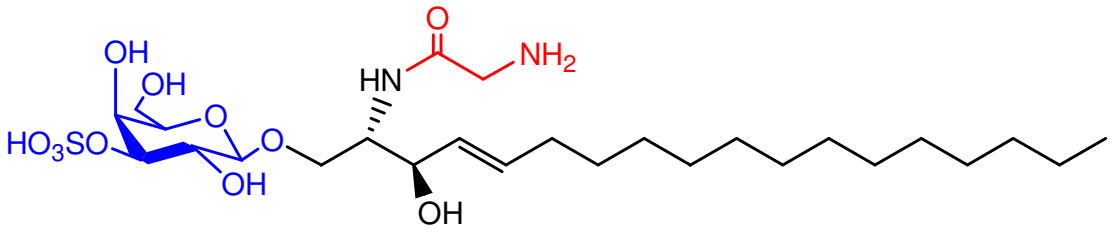
Sulfatides are 3-sulfated galactosylceramides that are found primarily in the central nervous system and are myelin specific sphingolipids. Over the last several decades sulfatides have been linked to many physiological functions and recently there has been a renewed interest in their role in diseases. Sulfatides are highly multifunctional glycolipids involved in the nervous system, diabetes, immune system, hemostasis/thrombosis, bacterial infection, and viral infection. By understanding the correlation between sulfatide's normal physiological functions and specific roles in disease, new diagnostic and therapeutic methods can be evaluated.

Abnormal sulfatide metabolism, such as in metachromatic leukodystrophy, can induce cell apoptosis due to endosome-mediated ceramide generation and the accumulation of cytotoxic levels of sulfatides in lysosomes.[1] Metachromatic leukodystrophy is an autosomal-recessive lysosomal storage disease caused by mutations in the arylsulfatase A (ARSA) gene leading to ARSA deficiency and causing sulfatide accumulation. Main symptoms of the disease are progressive demyelination, neurological dysfunction, and reduced life expectancy.[2] A comparative study analyzing sulfatide levels in newborn urine and dried blood spot samples concluded that urine, but not dried blood spots, were useful for screening newborns for metachromatic leukodystrophy.[3]

Sulfatides are highly dynamic glycosphingolipids with far reaching biological processes. A greater understanding of these functions in living systems will lead to the elucidation of associated diseases and the development of therapeutic treatments to various sulfatide associated diseases.

References:

1. X. Han et al. Endosomes and lysosomes play distinct roles in sulfatide-induced neuroblastoma apoptosis: potential mechanisms contributing to abnormal sulfatide metabolism in related neuronal diseases. Biochem. J. (2008) 410, 81
2. M. Manshadi et al., Four novel ARSA gene mutations with pathogenic impacts on metachromatic leukodystrophy: a bioinformatics approach to predict pathogenic mutations. Ther Clin Risk Manag. (2017) 13, 725-731
3. M. Barcenas et al. Quantification of sulfatides in dried blood and urine spots from metachromatic leukodystrophy patients by liquid chromatography/electrospray tandem mass spectrometry. Clinica Chimica Acta. 433, 39-43 (2014)



N-Glycinated *lyso*-sulfatide
(MS standard for *lyso*-sulfatide analysis)
Cat. 2092

Product Name	Catalog #	Purity
Sulfatides	1049	98+%
lyso-Sulfatide	1904	98+%
N-Glycinated *lyso*-sulfatide (MS standard for *lyso*)	2092	98+%
N-Acetyl-sulfatide (C2:0)	2076	98+%
N-Dodecanoyl-sulfatide (C12:0)	1938	98+%
N-Hexadecanoyl-sulfatide (C16:0)	1875	98+%
N-Heptadecanoyl-sulfatide (C17:0)	1934	98+%
N-Octadecanoyl-sulfatide (C18:0)	1932	98+%
N-Octadecenoyl-(*cis*-9)-sulfatide (C18:1)	1933	98+%
N-Nonadecanoyl-sulfatide (C19:0)	1935	98+%
N-Tetracosanoyl-sulfatide (C24:0)	1888	98+%
N-Tetracosenoyl-sulfatide (C24:1)	1931	98+%
N-Hexanoyl-biotin-sulfatide	2207	98+%
N-*omega*-CD_3-Octadecanoyl-sulfatide (deuterated)	1536	98+%
N-Dodecanoyl-NBD-sulfatide (fluorescent)	1632	98+%
N-Octadecanoyl-sulfated lactosylceramide	1540	98+%