

# MATREYA NEWSLETTER

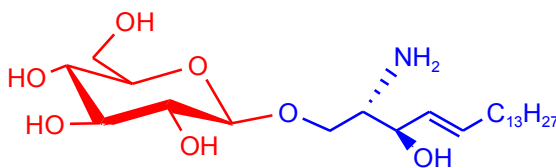
## FOR GLYCO/SPHINGOLIPID RESEARCH

### NOVEMBER 2015

Matreya appreciates your support over the past 25 YEARS. Due to your loyalty, Matreya has become a big success in the scientific community. We send a heartfelt THANK YOU to all of you, our customers, who have given us the chance to meet your needs over the years. We at Matreya enjoy working with you to help meet your research needs, supplying you with both standards and technical assistance to make your projects excel. We are looking forward to the next 25 years of being your preferred supplier. By working together, maybe we will find a cure for cancer, lysosomal storage disorders, and other diseases.



## Glucosylsphingosine – a Highly Sensitive and Specific Biomarker for Gaucher Disease



Catalog #2086

Lysosomal storage diseases (LSDs) are a heterogeneous group of disorders caused by lysosomal enzyme dysfunction<sup>(1)</sup>. Gaucher disease (GD) is the most common of these lysosomal storage disorders and has recently warranted much research due to the debilitating effects of excess lipid storage in Gaucher cells. A lack of activity in the lysosomal enzyme  $\beta$ -glucocerebrosidase causes accumulation of glucosylceramide, glucosylsphingosine and other glycosphingolipids. In lipid storage disorders it is very important to diagnose and treat patients as early as possible. One very effective method of diagnosis is the use of biomarkers.

Chitotriosidase is the most well-established biomarker for GD. However, it is not specific for GD and may give a false negative in a significant percentage of GD patients due to mutation. Chitotriosidase also reflects the changes in the course of the disease belatedly. Due to these limitations a more specific biomarker is needed for GD.

A. Rolfs et al. recently demonstrated that glucosylsphingosine can be used as a promising, reliable, and specific biomarker for GD. They evaluated the sensitivity and specificity of glucosylsphingosine with regard to healthy controls vs. Gaucher disease carriers vs. other LSDs control groups. Only GD patients displayed elevated levels of glucosylsphingosine higher than the pathological cut-off, verifying the specificity of glucosylsphingosine as a biomarker for GD. Glucosylsphingosine was monitored during enzyme replacement therapy and demonstrated a decrease in glucosylsphingosine over time<sup>(2)</sup>.

#### INSIDE THIS ISSUE

- Glucosylsphingosine for Gaucher Disease 1
- Sulfatides in Current Research 2
- Biotinylated Monosialoganglioside GM1 3
- CTH from Matreya 4

<u>Catalog #</u>	<u>Product Name</u>	<u>Amount</u>	<u>Purity</u>
2086	Glucosylsphingosine, synthetic	5 mg	98 <sup>+</sup> %
1306	Glucosylsphingosine, bovine buttermilk	5 mg	98 <sup>+</sup> %
1310	Glucosylsphingosine, plant	5 mg	98 <sup>+</sup> %

#### References:

1. Manger B., Z Rheumatol, 69:6 (2010) 527-38
2. A. Rolf et al., PLoS One 8:11 (2013) e79732

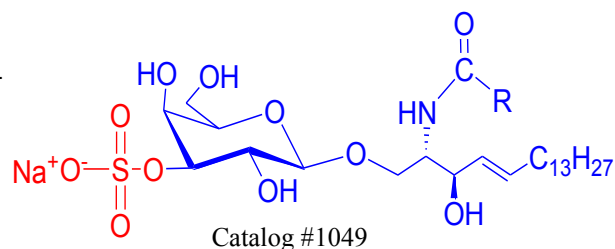
## Sulfatides in Current Research

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. By understanding the correlation between sulfatides, normal physiological functions, and specific diseases, new diagnostic and therapeutic methods can be evaluated.

Sulfatides derived from the brain and spinal cord have varying fatty acyl chains containing saturated, unsaturated, and 2-hydroxy acyl chains, the composition of which are vital in influencing function. They have been found to have many critical physiological roles, both inter- and intracellularly and are involved in numerous diseases, especially demyelinating diseases. Various infections, including influenza virus A and tuberculosis, have been demonstrated to be influenced by the actions of sulfatides<sup>(1)</sup>. Recently studies have implicated sulfatides in numerous inflammatory responses and there has been significant interest in its biological role towards CD1-restricted T cells. An important physiological role of sulfatides is its involvement in hemostasis and thrombosis by forming stable platelet aggregates<sup>(2,3)</sup>. 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<sup>(4)</sup>.

One of the most important areas of sulfatide research today is the study of its involvement in diseases and disorders. Sulfatide levels have been found to be elevated in ovarian cancer with the most prevalent sulfatide species detected via MALDI-TIMS being d18:1/C16:0, d18:1/C24:1 and d18:1/C24:0<sup>(5)</sup>. This elevation in sulfatides can be exploited as an ovarian cancer biomarker and as a possible therapeutic approach.

Due to its presence in the myelin sheath of nerves, it is not surprising that sulfatide metabolism has been implemented in many neural degenerative diseases. Sulfatide content precipitously drops in Alzheimers Disease and CNS sulfatide content has been found to be modulated by apolipoprotein E<sup>(6)</sup>. In multiple sclerosis the identity of the target antigen has long been unidentified but it was found that levels of antisulfatide antibodies were significantly higher in multiple sclerosis patients than in controls<sup>(7)</sup>. Sulfatide has also been demonstrated to be able to activate inflammatory responses as an (continued on page 3)



<u>Catalog #</u>	<u>Product Name</u>	<u>Amount</u>	<u>Purity</u>
1049	Sulfatides	50 mg	98 <sup>+</sup> %
1904	lyso-Sulfatide	1 mg	98 <sup>+</sup> %
2076	N-Acetyl-sulfatide (C2:0-sulfatide)	1 mg	98 <sup>+</sup> %
1938	N-Dodecanoyl-sulfatide (C12:0-sulfatide)	1 mg	98 <sup>+</sup> %
1875	N-Hexadecanoyl-sulfatide (C16:0-sulfatide)	1 mg	98 <sup>+</sup> %
1934	N-Heptadecanoyl-sulfatide (C17:0-sulfatide)	1 mg	98 <sup>+</sup> %
1932	N-Octadecanoyl-sulfatide (C18:0-sulfatide)	1 mg	98 <sup>+</sup> %
1933	N-Octadecenoyl-sulfatide (C18:1-sulfatide)	1 mg	98 <sup>+</sup> %
1935	N-Nonadecanoyl-sulfatide (C19:0-sulfatide)	1 mg	98 <sup>+</sup> %
1888	N-Tetracosanoyl-sulfatide (C24:0-sulfatide)	1 mg	98 <sup>+</sup> %
1931	N-Tetracosenoyl-sulfatide (C24:1-sulfatide)	1 mg	98 <sup>+</sup> %
1536	N-Octadecanoyl-D3-sulfatide (C18:0-D <sub>3</sub> -sulfatide)	1 mg	98 <sup>+</sup> %
1632	N-Dodecanoyl-NBD-sulfatide (C12:0-NBD-sulfatide)	100 µg	98 <sup>+</sup> %

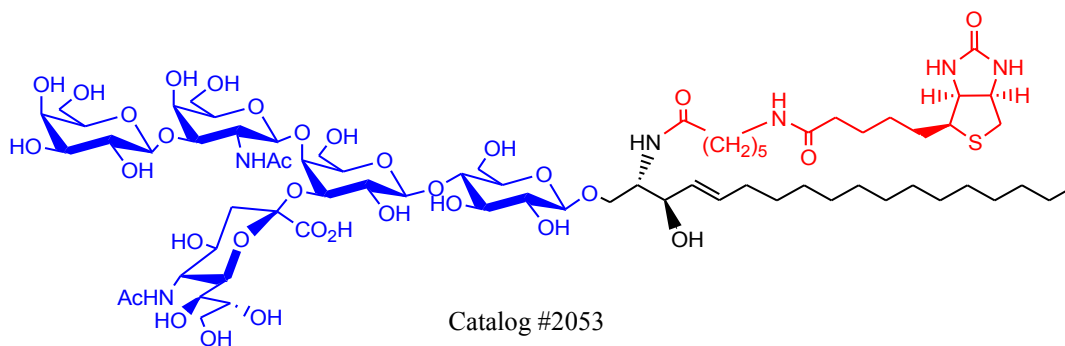
### References:

1. Y. Suzuki et al., *Biochem. J.*, 318 (1996) 389
2. P. Thiagarajan et al., *Arterioscler. Thromb. Vasc. Biol.*, 25 (2005) 258
3. M. Kyogashima, *Archives of Biochemistry and Biophysics*, 426:2 (2004) 157
4. X. Han et al. *Biochem. J.* 410 (2008) 81
5. A. Merrill Jr. et al., *Molecular Cancer* 9 (2010) 186
6. X. Han et al., *Journal of Neurochemistry* 106 (2008) 1275
7. A. Ilyas et al., *Journal of Neuroimmunology*, 139 (2003) 76
8. E. Park et al., *Journal of Immunology*, 181 (2008) 8077
9. Kumar et al., *Journal of Clinical Investigation*, 117 (2007) 2302
10. Kumar et al., *Neurochemical Research*, 32 (2007) 257
11. Kumar et al. *Gastroenterology*, 140:2 (2011) 646

endogenous stimulator in brain-resident immune cells<sup>(8)</sup>. Sulfatide derived from myelin binds promiscuously to several human CD1 molecules and to murine CD1d and is recognized by type II NKT cells. Among the sulfatides, *cis*-tetracosenoyl sulfatide is immunodominant and can either prevent or reverse antigen-induced experimental autoimmune encephalomyelitis in CD1d<sup>+/+</sup> mice<sup>(9)</sup>. It therefore may be able to reverse autoimmune demyelinating diseases in humans<sup>(10)</sup>.

In hepatic ischemic reperfusion injury subsets of NKT cells have opposing roles<sup>(11)</sup>. Type I NKT cells promote injury while sulfatide-reactive type II NKT cells protect against injury. CD1d activation of NKT cells is conserved from mice to humans, so strategies to modify these processes might be developed to treat patients with hepatic reperfusion injury.

## Biotin Labeled Ganglioside GM<sub>1</sub> for Enhanced Ganglioside Studies



Matreya's line of biotinylated sphingolipids are ideal for use in sphingolipid research, taking advantage of the strong and specific interaction of biotin with streptavidin/avidin. N-Hexanoyl-biotin-monosialoganglioside GM<sub>1</sub> contains a biotin label attached to the amine of the sphingosine moiety via a hexanoic acid linker which maintains the sphingolipid's natural properties. The biotin label allows for easy attachment of the sphingolipid to streptavidin/avidin proteins making it extremely useful for binding to substrates and for toxin detection<sup>(1)</sup>.

Gangliosides are acidic glycosphingolipids containing one or more sialic acids that generally form lipid rafts in the outer leaflet of the cell plasma membrane, especially in neuronal cells in the central nervous system<sup>(2,3)</sup>. They participate in many cellular activities including proliferation, differentiation, adhesion, signal transduction, cell-to-cell interactions, tumorigenesis, and metastasis<sup>(4)</sup>. The accumulation of gangliosides has been linked to several diseases including Tay-Sachs and Sandhoff disease, while an autoimmune response against gangliosides can lead to Guillain-Barre syndrome. Gangliosides act as receptors for several toxins and bacteria, accumulate in various tumors, and aid in many neuronal functions. They are therefore very important in therapeutic processes and have warranted much research. GM<sub>1</sub> stimulates neuronal sprouting and enhances the action of nerve growth factor (NGF) by directly and tightly associating with Trk, the high-affinity tyrosine kinase-type receptor for NGF. GM<sub>1</sub> has also been identified as the specific cell surface receptor for cholera toxin making it an important target for therapeutic interventions<sup>(5)</sup>.

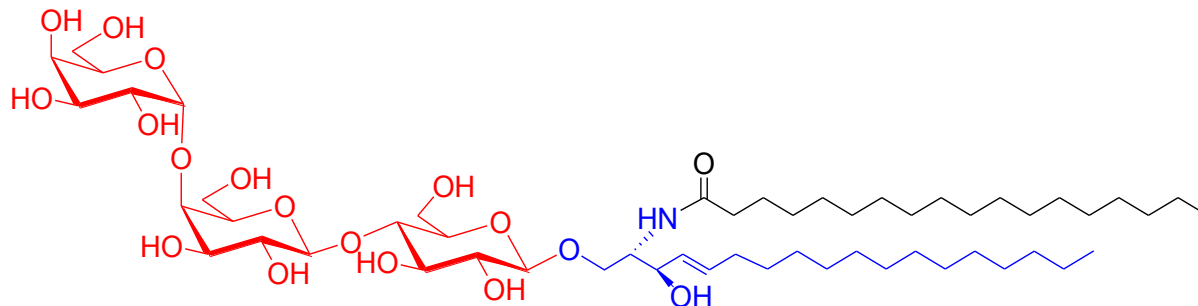
Pukin and coworkers used biotin labeled ganglioside analogs for *Escherichia coli* enterotoxin detection on streptavidin-coated ELISA plates<sup>(1)</sup>. Enterotoxigenic *Escherichia coli* is a pathogenic form of bacteria that is a serious threat to health and food safety around the world and the detection of enterotoxins is of critical importance in preventing food born diseases.

<u>Catalog #</u>	<u>Product Name</u>	<u>Amount</u>	<u>Purity</u>
2053	N-Hexanoyl-biotin-monosialoganglioside GM <sub>1</sub>	500 µg	98 <sup>+</sup> %

### References:

1. A. Pukin et al., *Org. Biomol. Chem.* 9:16 (2011) 5809-5815
2. L. Svennerholm, et al. (eds.), *Structure and Function of Gangliosides*, New York, Plenum, 1980
3. T. Kolter, R. Proia, K. Sandhoff, *J. Biol. Chem.* 277:29 (2002) 25859-25862
4. S. Birkle et al. *Biochimie*, 85 (2003) 455-463
5. C. E. Miller et al., *Biophysj.*, 86:6 (2004) 3700-3708

## Ceramide Trihexosides from Matreya



Catalog # 1067

Ceramide trihexoside (Gb<sub>3</sub>, CTH, globotriaosylceramide) is a very important cellular lipid that is involved in cell signaling, toxin receptor activation, and lysosomal storage disorders. Gb<sub>3</sub> plays an especially important role in Fabry disease where its accumulation causes a wide range of multisystemic symptoms affecting the heart, kidney and central nervous system.

Matreya has been offering very high purity natural Gb<sub>3</sub>, well-defined Gb<sub>3</sub>, and selected Gb<sub>3</sub> analogs for the past twenty years. We take great pride in offering these standards to aid our customer's research.

Matreya's commitment to superior product quality and unsurpassed technical service has been the foundational principle of our business. We designed our products based on customer needs, especially with regard to mass spectrometry and clinical analysis. We also tailor make our Gb<sub>3</sub> products for many of our customers in the field of therapeutic drug development. It is with great pride and passion that we offer the following Gb<sub>3</sub> standards.

<u>Catalog #</u>	<u>Product Name</u>	<u>Amount</u>	<u>Purity</u>
1067	Ceramide trihexosides	1 mg, 10 mg	98 <sup>+</sup> %
1513	Ceramide trihexosides (top spot)	500 µg	98 <sup>+</sup> %
1514	Ceramide trihexoside (bottom spot)	500 µg	98 <sup>+</sup> %
1520	lyso-Ceramide trihexoside	1 mg	98 <sup>+</sup> %
1530	N-Glycinated lyso-ceramide trihexoside	1 mg	98 <sup>+</sup> %
1528	N-Hexadecanoyl-ceramide trihexoside	500 µg	98 <sup>+</sup> %
1523	N-Heptadecanoyl-ceramide trihexoside	500 µg	98 <sup>+</sup> %
1529	N-Octadecanoyl-ceramide trihexoside	500 µg	98 <sup>+</sup> %
1524	N-Tricosanoyl-ceramide trihexoside	500 µg	98 <sup>+</sup> %
1631	N-Dodecanoyl-NBD-ceramide trihexoside	100 µg, 1 mg	98 <sup>+</sup> %
1537	N-Octadecanoyl-D <sub>3</sub> -ceramide trihexoside	500 µg	98 <sup>+</sup> %
1947	N-(1-Adamantaneacetyl)-ceramide trihexoside	1 mg	98 <sup>+</sup> %



### Free 2-Day Shipping!

Matreya is now offering free 2-day shipping on all continental U.S. orders of \$500 or more.