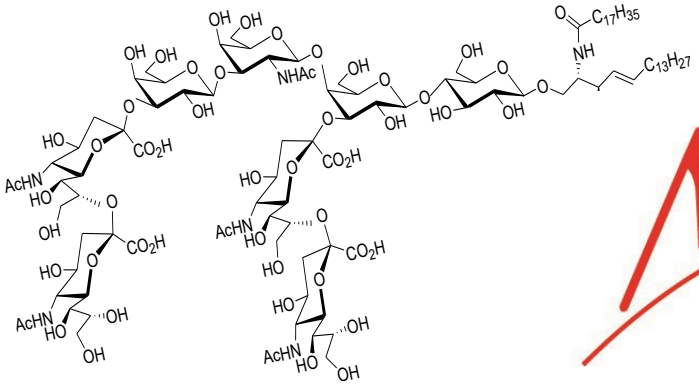
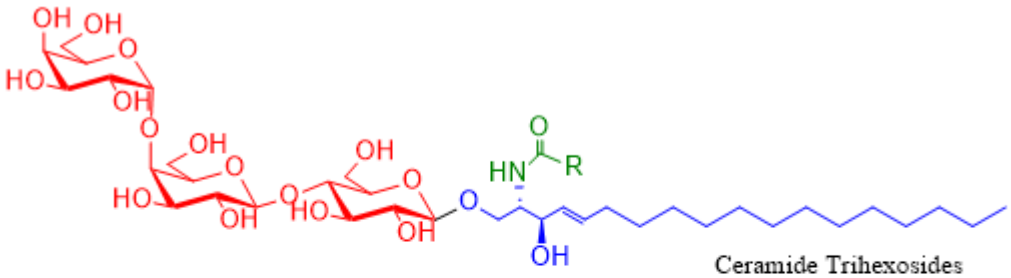


MATREYA LLC

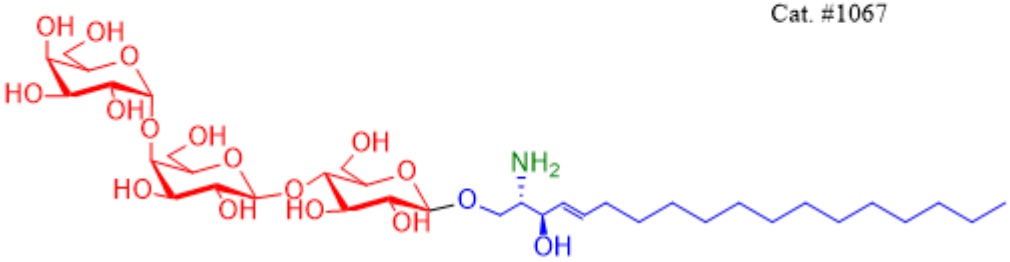
Lipids and Biochemicals

NEWSLETTER FOR GLYCO/SPHINGOLIPID RESEARCH
DECEMBER 2019

Gb_3 and *lyso*-Gb_3 and Fabry Disease



Ceramide Trihexosides
Cat. #1067



Lyso-Ceramide Trihexoides
Cat. #1520

Fabry disease is a multi-systemic x-linked disorder with variable prevalence ranging from 1:3000 to 1:117000 in newborn males.

Fabry disease is caused by deficiency in the alpha-galactosidase enzyme. Decrease in alpha-galactosidase enzyme leads to the storage of sphingolipids such as lyso-Gb3 and Gb3 and galabiosyl ceramide (Ga2) in organs, tissues, and biological fluids.

Enzyme replacement therapy (ERT) of the disease has been available since 2001. Several studies support the clinical benefit of ERT towards quality of life, disease progression, and stabilization of end organ structure and function.[1]

Thurberg[2] evaluated 48 Fabry patients on ERT and found good correlation of urinary Gb3 excretion normalized to creatine. However other studies indicated incomplete relationships between plasma and urinary Gb3 levels and disease manifestations. Recently, Aerts group[3] postulated Gb3 metabolite could play a role in Fabry pathogenesis and reposted the presence of lyso-Gb3 plasma. Elevated lyso-Gb3 were found in the plasma of Fabry patients and concentrations were reduced after ERT.

Inside This Issue

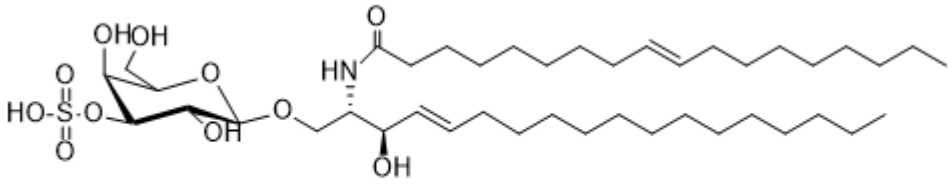
Plasma lyso-Gb3 excretion levels are an important indicator and correlated significantly into cerebrovascular white matter lesions in males and the left ventricle mass in females. Urinary lyso-Gb3 levels correct with mutation types, gender, and ERT status.

Product Name	Catalog #	Amount	Purity
Ceramide trihexoside (Globotriaosylceramide, Gb_3)	1067	1 mg	98+%
lyso-Ceramide trihexoside (Globotriaosylsphingosine, *lyso*-Gb3)	1520	1 mg	98+%
N-Glycinated *lyso*-ceramide trihexoside	1530	1 mg	98+%
N-Hexadecanoyl-ceramide trihexoside	1528	1 mg	98+%
N-Octadecanoyl-ceramide trihexoside	1529	500 µg	98+%
N-Tricosanoyl-ceramide trihexoside	1524	500 µg	98+%
N-*omega*-CD_3-Octadecanoyl-ceramide trihexoside	1537	500 µg	98+%
Ceramide trihexosides (top spot)	1513	500 µg	98+%
Ceramide trihexosides (bottom spot)	1514	500 µg	98+%
N-Dodecanoyl-NBD-ceramide trihexoside	1631	100 µg	98+%

Please visit www.matreya.com for a full list of ceramide trihexosides and other lipid standards.

References:
1. P. Lavoie, M. Boutin and C. Auray-Blais, Analytical Chem. 2013, 85, 1743-52.
2. Thurberg, B.L., Rennke, H., Colvin, R.B., Dikman, S., Gorden, R.E., Collins, A.B., Desnnick, R.J., O'Callaghan, M., Kidney Int. 2002, 62, 1933-46.
3. Aerts, J.M., Groener, J.E., Kuiper, D., Donker-Koopan, W.E., Strijland, A., Ottenhoff, R., Nan Roomen, C., Mirzan, M., Wijiburg, F., Linthorsk, G.E., Vedder, A.C., Romback, S.M., Cox-Brinkman, J., Somerhargu, P., Boot, R.G., Hollak, C.E., Brady, R.O., Poortheis, B.J. Proc. NaH. Acad. Sci, USA. 2008, 105, 2812, 2817.

The Biological Role of Sulfatides



N-Octadecenoyl-sulfatide (C18:1)
Cat# 1933

Sulfatides are 3'-sulfated galactosylceramides that are found primarily on oligodendrocytes, renal tubular cells, and some tumor cells. The most prominent role of sulfatides is in their involvement in nerve conduction and cell adhesion, although many other cellular functions have been observed. Sulfatides are able to induce intracellular signaling in neutrophils through a L-selectin dependent pathway and provide a binding site for human immunodeficiency virus, *Helicobacter pylori* and malaria sporozites. In the brain and spinal cord sulfatides have different molecular species of varying fatty acyl chains containing saturated, unsaturated, and 2-hydroxy fatty acids, the composition of which are vital in influencing their function. Over the last several decades sulfatides have been linked to many physiological processes and recently there has been a renewed interest in their role in diseases.

Demyelintating Diseases:

Both sulfatide and its metabolic precursor galactosylcermide are present in high concentrations in multilamellar layers of the myelin surrounding the axons of neuronal cells where they are involved in nerve conduction. A production of anti-sulfatide antibodies in the cerebrospinal fluid, leading to a deficiency in sulfatides, may be a cause of degeneration of the myelin sheath, leading to multiple sclerosis and other demyelinating diseases[(1)].

Platelet Aggregation:

Sulfatides interact with several cell adhesion molecules such as laminin, thrombospondin, von Willibrand factor, and selectin[2]. They have recently been identified as a major ligand for P-selectin in platelet adhesion aggregation which is necessary for the formation of stable platelet aggregates. Platelets expressing sulfatides were found to adhere to P-selectin while platelets expressing P-selectin adhered to sulfatides. When sulfatide presence was masked by forming sulfatide micelles or introducing sulfatide-binding recombinant malaria circumsporozoite protein the adhesion was inhibited. The role of sulfatide/P-selectin interaction may play a significant role in hemostasis and thrombosis[3].

Leukotriene Synthesis:

In adherent human polymorphonuclear leukocytes sulfatides have been found to suppress leukotriene synthesis by directly inhibiting 5-lipoxygenase and impeding its translocation to the nuclear envelope. The mechanism for this inhibition may be due to sulfatide causing a redistribution of cholesterol, increasing its abundance at the uropod region[4].

Ovarian Tumors:

Cancerous cells are known to present unusually high levels of specific molecules, especially glycosphingolipids. Sulfatides have now been identified as being present in elevated levels in patients having advanced ovarian tumors. The elevated levels of sulfatide in these cells may play a role in the pathogenesis of ovarian cancer. These elevated sulfatide levels can be used as a biomarker to predict the presence of advanced stage ovarian cancer even when the patient otherwise appears to be in the early stage of the disease[5].

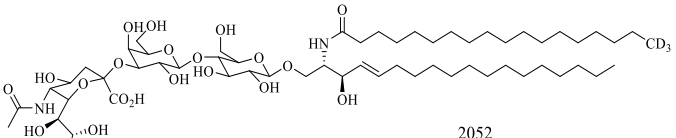
Available Sulfatides	Catalog #	Amount	Purity
Sulfatides (bovine)	1049	1 mg	98+%
lyso-sulfatide	1904	1 mg	98+%
N-Acetyl-sulfatide (C2:0)	1530	1 mg	98+%
N-Hexadecanoyl-sulfatide (C16:0)	1528	1 mg	98+%
N-Octadecanoyl-sulfatide (C18:0)	1529	500 µg	98+%
N-Octadecenoyl-sulfatide (C18:1)	1933	1 mg	98+%
N-Tetracosanoyl-sulfatide (C24:0)	1888	1 mg	98+%
N-Tetracosenoyl-sulfatide (C24:1)	1931	1 mg	98+%
N-Octadecanoyl-D_3-sulfatide (C18:0 D_3)	1536	1 mg	98+%
N-Dodecanoyl-NBD-sulfatide (C12:0 NBD)	1632	100 µg	98+%

References:
1. Halder C., Jahng A., Maricic I., and Kumar V., Neurochemical Research, 32(2): 257-262 (2007)
2. Borthakler G., Cruz M., Dong J., McIntire L., Li F., Lopez J., and Thiagarajan P., J. Thromb. Haemost. 1(6): 1288-1295 (2003)
3. Merten, M. and Thiagarajan, P., Circulation, 104: 2955-2960 (2001)
4. Grishina Z., Pushkareva M., Pletjushkina O., Reiser G., Peters-Golden M., Sud'ina G., Int. J. Biochem. Cell Biol. 40(1): 110-124 2008
5. Makhlouf A., Fathalla M., Zakhary M. and Makarem M., Int J. Gynecol Cancer 14(1): 89-93 (2004)

Deuterated Lipids

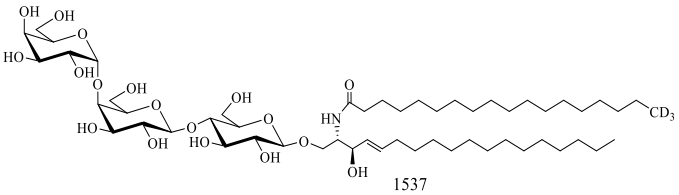
Matreya offers a variety of high purity deuterated labelled glycosphingolipids. These compounds are made by several steps and rigorous purifications. These are ideal as mass spectrometric standards and can be used as biomarkers.

If you need any other deuterium labeled compounds please do not hesitate to contact us.



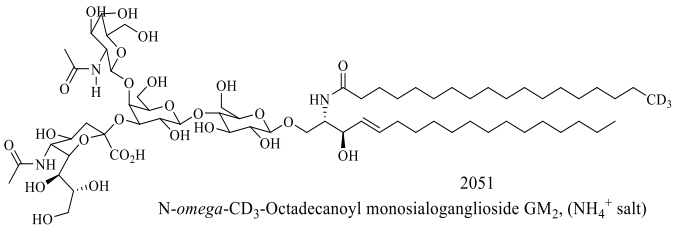
2052

N-*omega*-CD_3-Octadecanoyl monosialoganglioside GM_3, (NH_4^+ salt)



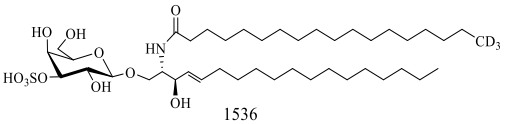
1537

N-*omega*-CD_3-Octadecanoyl-ceramide trihexoside



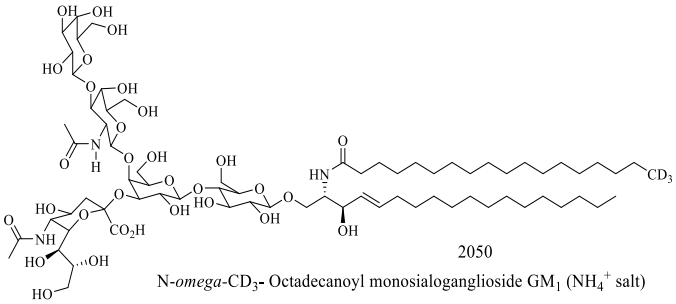
2051

N-*omega*-CD_3-Octadecanoyl monosialoganglioside GM_2, (NH_4^+ salt)



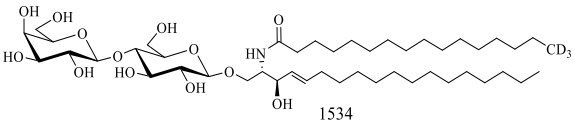
1536

N-*omega*-CD_3-Octadecanoyl-sulfatide



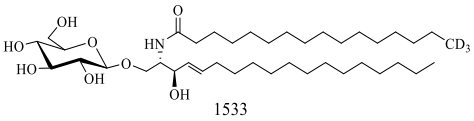
2050

N-*omega*-CD_3- Octadecanoyl monosialoganglioside GM_1 (NH_4^+ salt)



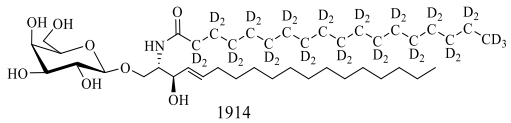
1534

N-*omega*-CD_3-Hexadecanoyl-lactosylceramide



1533

N-*omega*-CD_3-Hexadecanoyl-glucopsychosine



1914

N-Octadecanoyl-D_{35}-psychosine, (perdeuterated, C18:0 fatty acid)

Product Name	Catalog #	Amount	Purity
N-Octadecanoyl-D_{35}-galactopsychosine	1914	5 mg	98+%
N-Hexadecanoyl-D_3-glucopsychosine	1533	1 mg	98+%
N-Hexadecanoyl-D_3-lactosylceramide	1534	1 mg	98+%
N-Octadecanoyl-D_3-sulfatide	1536	1 mg	98+%
N-Octadecanoyl-D_3-ceramide trihexoside	1537	500 µg	98+%
N-Octadecanoyl-D_3- monosialoganglioside GM_1	2050	500 µg	98+%
N-Octadecanoyl-D_3- monosialoganglioside GM_2	2051	250 µg	98+%
N-Octadecanoyl-D_3- monosialoganglioside GM_3	2052	250 µg	98+%

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