

PRODUCT INFORMATION



Lyso-Globotriaosylceramide (d18:1)

Item No. 24873

CAS Registry No.: 126550-86-5

Formal Name: (2S,3R,4E)-2-amino-3-hydroxy-4-octadecen-1-yl O- α -D-galactopyranosyl-(1 \rightarrow 4)-O- β -D-galactopyranosyl-(1 \rightarrow 4)- β -D-glucopyranoside

Synonyms: Globotriaosylsphingosine (d18:1), Lyso-Ceramide Trihexoside, Lyso-Gb₃

MF: C₃₆H₆₇NO₁₇

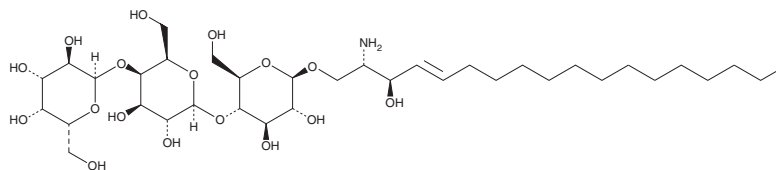
FW: 785.9

Purity: \geq 98%

Supplied as: A solid

Storage: -20°C

Stability: \geq 4 years



Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.

Laboratory Procedures

Lyso-globotriaosylceramide (d18:1) is supplied as a solid. A stock solution may be made by dissolving the lyso-globotriaosylceramide (d18:1) in the solvent of choice. Lyso-globotriaosylceramide (d18:1) is soluble in a 4:3:1 solution of chloroform:methanol:water.

Description

Lyso-globotriaosylceramide is a form of globotriaosylceramide that is lacking the fatty acyl group. It binds to Shiga toxin 1 (Stx1) in the presence of cholesterol and phosphatidylcholine but does not bind Stx2.¹ It also reduces viability and aggregation of human neutrophils induced by phorbol 12-myristate 13-acetate (PMA; Item No. 10008014) when used at concentrations of 50 and 1 μ M, respectively.² Lyso-globotriaosylceramide accumulates in the brain, heart, kidney, liver, lung, and spleen in a mouse model of Fabry disease, a lysosomal storage disorder characterized by a deficiency in the enzyme α -galactosidase A.³ It also accumulates in the urine, kidney, and plasma of patients with Fabry disease.⁴ Lyso-globotriaosylceramide levels decrease in response to administration of the α -galactosidase inhibitor 1-deoxygalactonojirimycin (migalastat; Item No. 17179) in a transgenic mouse model of Fabry disease. Decreases in plasma and urine concentrations of lyso-globotriaosylceramide have been used as a biomarker for efficacy of enzyme replacement therapy (ERT) and other therapies in the treatment of Fabry disease.

References

- Gallegos, K.M., Conrady, D.G., Karve, S.S., *et al.* Shiga toxin binding to glycolipids and glycans. *PLoS One* **7**(2), e30368 (2012).
- Fiore, S., Nicolaou, K.C., Caulfield, T., *et al.* Evaluation of synthetic sphingosine, lysosphingolipids and glycosphingolipids as inhibitors of functional responses of human neutrophils. *Biochem. J.* **266**(1), 25-31 (1990).
- Kamani, M.A., Provençal, P., Boutin, M., *et al.* Glycosphingolipid storage in Fabry mice extends beyond globotriaosylceramide and is affected by ABCB1 depletion. *Future Sci. OA.* **2**(4), FS0147, (2016).
- Young-Gqamana, B., Brignol, N., Chang-H.H., *et al.* Migalastat HCl reduces globotriaosylsphingosine (lyso-Gb₃) in Fabry transgenic mice and in the plasma of Fabry patients. *PLoS One* **8**(3), e57631 (2013).

WARNING

THIS PRODUCT IS FOR RESEARCH ONLY - NOT FOR HUMAN OR VETERINARY DIAGNOSTIC OR THERAPEUTIC USE.

SAFETY DATA

This material should be considered hazardous until further information becomes available. Do not ingest, inhale, get in eyes, on skin, or on clothing. Wash thoroughly after handling. Before use, the user must review the complete Safety Data Sheet, which has been sent via email to your institution.

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