

PRODUCT DATA SHEET

lyso-Ceramide trihexoside

Catalog No: 1520

Common Name: *lyso*-CTH; *lyso*-
Globotriaosylsphingosine

Source: semisynthetic, porcine RBC

Solubility: chloroform/methanol/DI water
(2:1:0.1) ; DMSO

CAS No: 126550-86-5

Molecular Formula: C₃₆H₆₇NO₁₇

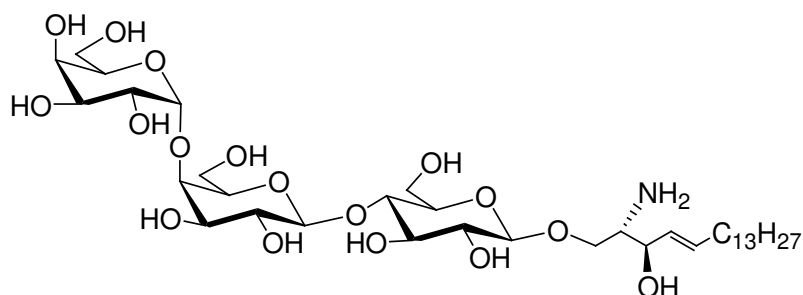
Molecular Weight: 786

Storage: -20°C

Purity: TLC > 98%; identity confirmed by MS

TLC System: chloroform/methanol/DI water/
2.5N ammonium hydroxide
(60:40:7:3 by Vol.)

Appearance: solid



Application notes:

lyso-Ceramide trihexoside contains a free amine linkage enabling well-defined ceramide trihexosides to be produced. Ceramide trihexoside is involved in cellular signaling and has been identified as a receptor for various toxins including shiga toxins and shiga-like toxins.¹ Some toxins, such as verotoxins from *Escherichia coli*, require specific fatty acids on the ceramide portion of CTH to show affinity in binding. An accumulation of CTH in the cellular membrane due to a lack of alpha-galactosidase to convert it into lactosyl ceramide results in Fabry disease.² It can be used as an excellent standard for the identification of CTH in Fabry disease by HPLC³ and mass spectrometry.⁴ In contrast to Fabry disease, a deficiency of CTH due to mutations in the gene sequence leads to the P^k Blood Group Phenotype. It appears that under certain conditions CTH can enhance anticoagulant activity. CTH has also been studied as a tool to investigate lymphocyte activation.⁵

Selected References:

1. M. Jacewicz, H. Clausen, E. Nudelman, A. Donohue-Rolfe, G. Keusch "Pathogenesis of shigella diarrhea. XI. Isolation of a shigella toxin-binding glycolipid from rabbit jejunum and HeLa cells and its identification as globotriaosylceramide" *J Exp Med.*, Vol.163:6 pp.1391-1404, 1986
2. S. Bekri, O. Lidove, R. Jaussaud, B. Knebelmann, F. Barbey "The role of ceramide trihexoside (globotriaosylceramide) in the diagnosis and follow-up of the efficacy of treatment of Fabry disease: a review of the literature. Cardiovasc Hematol Agents" *Med Chem* Vol. 4:4 pp. 289-297, 2006
3. J. Groener, et al. "HPLC for simultaneous quantification of total ceramide, glucosylceramide, and ceramide trihexoside concentrations in plasma" *Clin Chem.* Vol. 53:4 pp.742-747, 2007
4. K. Mills, A. Johnson, B. Winchester "Synthesis of novel internal standards for the quantitative determination of plasma ceramide trihexoside in Fabry disease by tandem mass spectrometry" *FEBS Lett.*, Vol. 515 pp. 171-176, 2002
5. C. Menge et al. "Globotriaosylceramide (Gb(3)/CD77) is synthesized and surface expressed by bovine lymphocytes upon activation in vitro" *Vet Immunol Immunopathol.*, Vol. 83 pp.19-36, 2001

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