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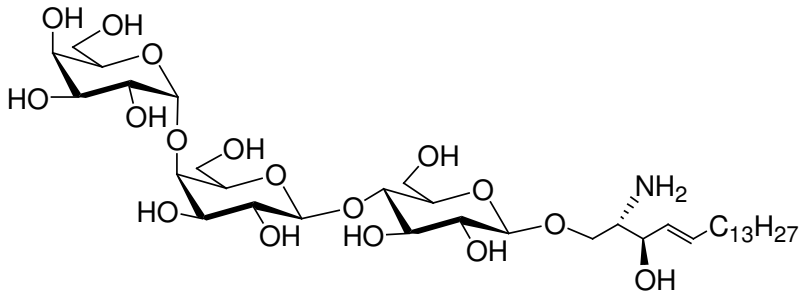
PRODUCT DATA SHEET

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)
CAS No: 126550-86-5

Molecular Formula: $C_{36}H_{67}NO_{17}$
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 veratoxins 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 HPLC[3] 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*