

PRODUCT DATA SHEET

製品名: lyso-Ceramide trihexosides

カタログ番号: 1520

別名: lyso-CTH; lyso-Gb3; lyso-Globotriaosylceramide etc.

由来: Semisynthetic, porcine RBC

溶解度: Chloroform/Methanol/Water (2: 1: 0.1)

CAS 番号: 126550-86-5

分子式: C₆₀H₆₇NO₁₇

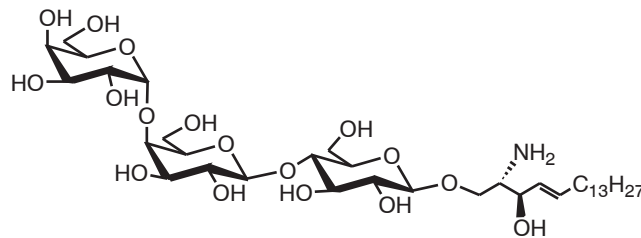
分子量: 786

保存温度: -20 °C

純度: TLC >98%

TLC 溶媒: Chloroform/ Methanol/ Water/
2.5N Ammonium hydroxide
(60: 30: 5: 3 by Vol.)

形状: Solid



Application notes:

Lyso-Ceramide trihexoside contains a free amine linkage enabling well-defined ceramide trihexosides to be produced. Ceramide trihexoside (CTH) is a glycosphingolipid found mostly in mammalian cell membranes. It 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 membranes due to a lack of alpha-galactosidase to convert it into lactosyl ceramide results in Fabry disease.² This product can be used as an excellent standard for the identification of CTH in Fabry disease by HPLC³ and mass spectrometry.⁴ An inability to convert CTH to globoside 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

アプリケーションノート

リソセラミドトリヘキシドは、自由なアミン結合を有しているため、アミド結合によるセラミドトリヘキシドの生成が可能です。セラミドトリヘキシドは細胞シグナル伝達に関与し、志賀毒素や志賀様毒素を含む様々な毒素の受容体として同定されています。幾つかの毒素、例えば大腸菌のペロ毒素は、結合親和性を示すために CTH のセラミド部分に特定の脂肪酸を必要とします。CTH をラクチルセラミドに変換する α-ガラクトシダーゼが欠損・活性低下すると、細胞膜に CTH が蓄積しファブリー病 (Fabry's disease) を発症します。本製品は、HPLC や質量分析によるファブリー病の CTH 同定のために、非常に優れた標準品として用いることが可能です。遺伝子配列の突然変異により CTH のグロボシドへの変換ができないことが原因で P^k 抗原群の表現型となります。ある環境下では CTH は抗凝活性を高めることができます。CTH はリンパ球の活性を調べるためのツールとして研究されることもあります。

Selected References:

1. S. Ashkenazi and T. G. Cleary, "Rapid method to detect shiga toxin and shiga-like toxin I based on binding to globotriaosyl ceramide (Gb3), their natural receptor." *J Clin Microbio.* June; 27(6): 1145-1150, 1989
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* 4 (4): 289-97, October 2006
3. J. E. Groener, B. J. Poorthuis, S. Kuiper, M. T. Helmond, C. E. Hollak, J. M. Aerts. "HPLC for simultaneous quantification of total ceramide, glycosylceramide, and ceramide trihexoside concentrations in plasma." *Clin Chem.*, Apr;53(4):742-7, 2007. Epub Mar 1 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.*, Mar 27;515(1-3):171-6, 2002
5. C. Menge, I. Stamm, M. Wuhler, R. Geyer, L. H. Wieler, G. Baljer. "Globotriaosylceramide (Gb3)/CD77 is synthesized and surface expressed by bovine lymphocytes upon activation in vitro." *Vet Immunol Immunopathol.*, Nov;83(1-2):19-36, 2001

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