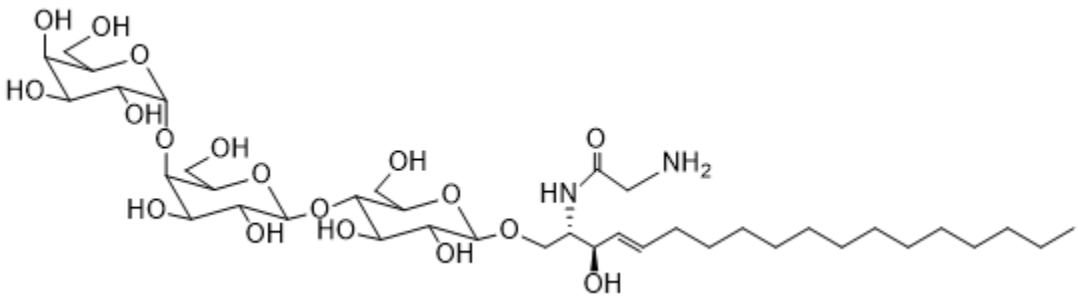
PRODUCT DATA SHEET

N-Glycinated *lyso*-ceramide trihexoside

Catalog number: 1530
Synonyms: N-Glycinated globotriaosylsphingosine; Internal standard for globotriaosylsphingosine
Source: semisynthetic, porcine RBC
Solubility: chloroform/methanol/DI water 2:1:0.1, DMF
CAS number: 1360882-59-2

Molecular Formula: $C_{38}H_{70}N_2O_{18}$
Molecular Weight: 843
Storage: -20°C
Purity: 98+%; identity confirmed by MS
TLC System: chloroform/methanol/DI water/2.5 N ammonium hydroxide, 60:40:7:3
Appearance: solid



Application Notes:

N-Glycinated *lyso*-ceramide trihexoside is an analogue of the important biomolecule *lyso*-ceramide trihexoside (globotriaosylsphingosine, *lyso*-Gb3). It is ideal for use as an internal standard in the extraction and mass spectrometry (MS) analysis of *lyso*-ceramide trihexoside.(1) The free amine group gives this product very similar physical characteristics to the natural *lyso*-ceramide trihexoside while the glycine adds an additional 57 units to the molecule making it easy to detect by MS. *lyso*-Ceramide trihexoside and the acylated ceramide trihexoside (globotriaosylceramide, Gb3) are important biomarkers for the lysosomal storage disorder Fabry disease.(2) Fabry disease is characterized by a deficiency in the enzyme α-galactosidase, resulting in an accumulation of ceramide trihexoside and *lyso*-ceramide trihexoside.(3) Early detection and treatment of this disease is critical to prevent damage to various organs.

Selected References:

1. R. Krüger et al. Quantification of the Fabry marker lysoGb3 in human plasma by tandem mass spectrometry. *Journal of Chromatography B.*, Vol. 883-884, pp. 128-135, 2012
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. C. Auray-Blais et al. How well does urinary lyso-Gb3 function as a biomarker in Fabry disease. *Clin. Chim. Acta*, Vol. 411(23-24) pp. 1906-1914, 2010

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