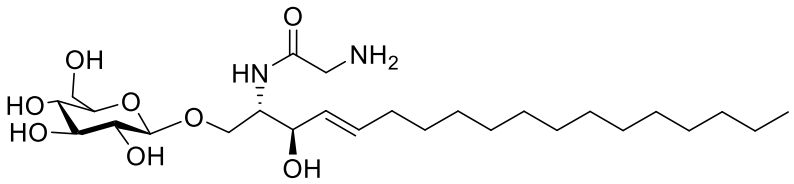
PRODUCT DATA SHEET

N-Glycinated glucosylsphingosine

Catalog No: 2089
Common Name: N-Glycine glucopsychosine; N-Glycinated 1-*beta*-D-*lyso*-glucosylceramide
Source: synthetic
Solubility: chloroform/methanol 8:2; DMSO; DMF
CAS No: N/A

Molecular Formula: $C_{26}H_{50}N_2O_8$
Molecular Weight: 519
Storage: -20°C
Purity: TLC > 98%; identity confirmed by MS
TLC System: chloroform/methanol/DI water/ ammonium hydroxide (65:25:3:2 by vol.)
Appearance: solid



Application Notes:

N-Glycinated glucosylsphingosine is an analogue of the important biomolecule glucosylsphingosine. It is ideal for use as an internal standard in the extraction and mass spectrometry analysis of glucosylsphingosine from natural samples.(1) The free amine group gives this product very similar physical characteristics to the natural glycolipid while the glycine adds an additional 57 units to the molecule making it easy to detect by MS.

Glucosylceramide and glucosylsphingosine are important biological species that are the precursors for many complex glycosphingolipids. These lipids are involved in critical cellular functions such as cellular proliferation, differentiation, adhesion, signal transduction, cell-to-cell interactions, tumorigenesis, and metastasis. One of the most important roles of glucosylsphingosine is as a biomarker for the lysosomal storage disorder Gaucher disease.

Gaucher disease is characterized by an accumulation of glucocerebroside due to a deficiency in the enzyme glucocerebrosidase and it has now been found that glucosylsphingosine also accumulates in this disease.(2) this accumulation of glucopsychosine contributes to neuronal dysfunction and destruction in patients with neuronopathic Gaucher disease(3) and it has been found to be a potent inhibitor of glucocerebrosidase. At least some instances of Gaucher disease also have a deficiency in the activity of glucosylsphingosine *beta*-glucosidase, the enzyme responsible for cleaving off the glucose of glucopsychosine and glucocerebroside. Like glucocerebroside and galactocerebroside, glucosylsphingosine can increase Ca_2^+ mobilization from intracellular stores although it uses a different mechanism.(4)

Selected References:

1. R. Krüger et al. Quantification of the Fabry marker *lyso*Gb_3 in human plasma by tandem mass spectrometry. *Journal of Chromatography B.*, Vol. 883-884, pp. 128-135, 2012
2. E. Orvisky et al. "Glucosylsphingosine accumulation in tissues from patients with Gaucher disease: correlation with phenotype and genotype" *Molecular genetics and metabolism*, Vol. 76(4) pp. 262-270, *2002*
3. R. Brady et al. "Toxicity of glucosylsphingosine (glucopsychosine) to cultured neuronal cells: a model system for assessing neuronal damage in Gaucher disease type 2 and 3" *Neurobiology of Disease*, Vol. 14(3) pp. 595-601, *2003*
4. E. Loyd-Evans et al. "Glucosylceramide and Glucosylsphingosine Modulate Calcium Mobilization from Brain Microsomes via Different Mechanisms" *The Journal of Biological Chemistry*, Vol. 278 pp. 23594-23599, *2003*