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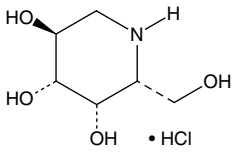
Product Information

1-Deoxygalactonojirimycin (hydrochloride)

Item No. 17179

CAS Registry No.: 75172-81-5
Formal Name: (2R,3S,4R,5S)-2-(hydroxymethyl)-3,4,5-piperidinetriol, monohydrochloride
Synonyms: DGJ, Migalastat
MF: $C_6H_{13}NO_4 \cdot HCl$
FW: 199.6
Purity: ≥98%
Stability: ≥2 years at -20°C
Supplied as: A crystalline solid
UV/Vis.: λ_{max}: 314 nm



Laboratory Procedures

For long term storage, we suggest that 1-deoxygalactonojirimycin (hydrochloride) be stored as supplied at -20°C. It should be stable for at least two years.

1-Deoxygalactonojirimycin (hydrochloride) is supplied as a crystalline solid. A stock solution may be made by dissolving the 1-deoxygalactonojirimycin (hydrochloride) in the solvent of choice. 1-Deoxygalactonojirimycin (hydrochloride) is soluble in DMSO, which should be purged with an inert gas. The solubility of 1-deoxygalactonojirimycin (hydrochloride) in this solvent is approximately 10 mg/ml.

Further dilutions of the stock solution into aqueous buffers or isotonic saline should be made prior to performing biological experiments. Ensure that the residual amount of organic solvent is insignificant, since organic solvents may have physiological effects at low concentrations. Organic solvent-free aqueous solutions of 1-deoxygalactonojirimycin (hydrochloride) can be prepared by directly dissolving the crystalline solid in aqueous buffers. The solubility of 1-deoxygalactonojirimycin (hydrochloride) in PBS, pH 7.2, is approximately 1 mg/ml. We do not recommend storing the aqueous solution for more than one day.

Fabry disease is an X-linked hereditary lysosomal storage disorder caused by mutations in the α-galactosidase gene that commonly lead to enzyme instability, misfolding, and degradation.[1,2] 1-Deoxygalactonojirimycin is a competititve inhibitor of α-galactosidase (IC_{50} = 40 nM).[3] At subinhibitory concentrations, 1-Deoxygalactonojirimycin binds to α-galactosidase and chaperones unstable enzyme variants through the endoplasmic reticulum, allowing its movement into lysosomes.[1,4] Pharmacological chaperones, including 1-Deoxygalactonojirimycin , are used to promote lysosomal delivery of unstable proteins in lysosomal storage disorders, like Fabry disease.[1]

References

1. Siekierska, A., De Baets, G., Ruemers, J., *et al.* α-Galactosidase aggregation is a determinant of pharmacological chaperone efficacy on Fabry disease mutants. *J. Biol. Chem.* **287(34)**, 28386-28397 (2012).
2. Lukas, J., Giese, A.-K., Markoff, A., *et al.* Functional characterisation of α-galactosidase a mutations as a basis for a new classification system in fabry disease. *PLoS One* **9(8)**, 1-10 (2013).
3. Asano, N., Ishii, S., Kizu, H., *et al. In vitro* inhibition and intracellular enhancement of lysosomal α-galactosidase A activity in Fabry lymphoblasts by 1-deoxygalactonojirimycin and its derivatives. *Eur. J. Biochem.* **267(13)**, 4179-4186 (2000).
4. Ishii, S., Chang, H.-H., Yoshioka, H., *et al.* Preclinical efficacy and safety of 1-deoxygalactonojirimycin in mice for Fabry disease. *J. Pharmacol. Exp. Ther.* **328(3)**, 723-731 (2009).

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