

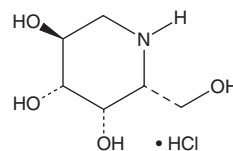
PRODUCT INFORMATION



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: $\geq 98\%$
UV/Vis.: λ_{max} : 314 nm
Supplied as: A crystalline solid
Storage: $-20^\circ C$
Stability: ≥ 4 years



Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.

Laboratory Procedures

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, which should be purged with an inert gas. 1-Deoxygalactonojirimycin (hydrochloride) is soluble in the organic solvent DMSO at a concentration of 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.

Description

1-Deoxygalactonojirimycin is a competitive inhibitor of α -galactosidase ($IC_{50} = 40$ nM).¹ At subinhibitory concentrations, 1-Deoxygalactonojirimycin binds to α -galactosidase and chaperones unstable enzyme variants through the endoplasmic reticulum, allowing its movement into lysosomes.^{2,3} Pharmacological chaperones, including 1-Deoxygalactonojirimycin, are used to promote lysosomal delivery of unstable proteins in lysosomal storage disorders, like Fabry disease.³

References

1. 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).
2. 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).
3. 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).

WARNING

THIS PRODUCT IS FOR RESEARCH ONLY - NOT FOR HUMAN OR VETERINARY DIAGNOSTIC OR THERAPEUTIC USE.

SAFETY DATA

This material should be considered hazardous until further information becomes available. Do not ingest, inhale, get in eyes, on skin, or on clothing. Wash thoroughly after handling. Before use, the user must review the [complete](#) Safety Data Sheet, which has been sent via email to your institution.

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