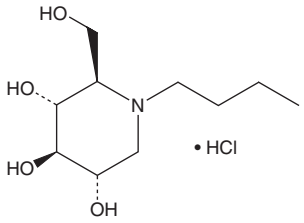
PRODUCT INFORMATION

N-Butyldeoxynojirimycin (hydrochloride)

Item No. 21065

CAS Registry No.:	210110-90-0
Formal Name:	1-butyl-2R-(hydroxymethyl)-3R,4R,5S-piperidinetriol, monohydrochloride
Synonym:	Miglustat, NB-DNJ
MF:	$C_{10}H_{21}NO_4$ • HCl
FW:	255.7
Purity:	≥98%
Supplied as:	A crystalline solid
Storage:	-20°C
Stability:	≥4 years



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

Laboratory Procedures

N-Butyldeoxynojirimycin (NB-DNJ) (hydrochloride) is supplied as a crystalline solid. A stock solution may be made by dissolving the NB-DNJ (hydrochloride) in the solvent of choice which should be purged with an inert gas, NB-DNJ (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 NB-DNJ (hydrochloride) can be prepared by directly dissolving the crystalline solid in aqueous buffers. The solubility of NB-DNJ (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

NB-DNJ is an iminosugar that inhibits UDP-glucose ceramide glucosyltransferase and β-glucosidase 2 (IC_{50}s = 32 and 81 µM, respectively, for the rat recombinant enzymes).[1] It also increases the activity of wild-type acid β-glucosidase, as well as the S364R, N370S, V15M, or M123T mutants, in COS-7 cells expressing the human enzymes when used at a concentration of 10 µM.[2] NB-DNJ also inhibits HIV-1 and HIV-2 infection of peripheral blood mononuclear cells (PBMCs; IC_{50}s = 282 and 211 µM, respectively).[3] Formulations containing NB-DNJ have been used in the treatment of Gaucher disease, an inborn error of metabolism characterized as a lysosomal storage disorder resulting from substantial deficiency of β-glucosidase.

References

1. Lee, J.C., Francis, S., Dutta, D., *et al.* Synthesis and evaluation of eight- and four-membered iminosugar analogues as inhibitors of testicular ceramide-specific glucosyltransferase, testicular β-glucosidase 2, and other glycosidases. *J. Org. Chem.* **77(7)**, 3082-3098 (2012).
2. Alfonso, P., Pampín, S., Estrada, J., *et al.* Miglustat (NB-DNJ) works as a chaperone for mutated acid β-glucosidase in cells transfected with several Gaucher disease mutations. *Blood Cells Mol. Dis.* **35(2)**, 268-276 (2005).
3. Pollock, S., Dwek, R.A., Burton, D.R., *et al.* N-Butyldeoxynojirimycin is a broadly effective anti-HIV therapy significantly enhanced by targeted liposome delivery. *AIDS* **22(15)**, 1961-1969 (2008).

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