

Catalog # 10-2413

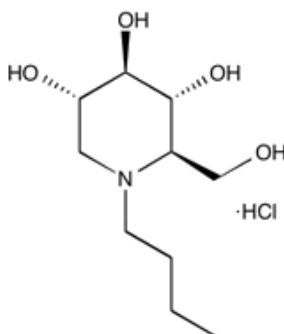
Miglustat HCl

CAS# 210110-90-0

N-Butyldeoxynojirimycin HCl

NB-DNJ

Lot # X101478



Orally active ceramide-specific glycosyltransferase and α -glucosidase I and II inhibitor.¹ Rescues trafficking-deficient F508del-CFTR in human airway epithelial cells via inhibition of ER α -glucosidases I and II.² Pharmacological chaperone for glucocerebrosidase degradation.³ Clinically useful agent for Gaucher disease type 1.⁴ Stabilizes neurological disorders in Niemann-Pick disease type C.⁵

- 1) Platt et al. (1994), *N-butyldeoxynojirimycin is a novel inhibitor of glycolipid biosynthesis*; J. Biol. Chem., **269** 8362
- 2) Noel et al. (2008), *Parallel improvement of sodium and chloride transport defects by miglustat (n-butyldeoxynojirimycin) in cystic fibrosis*; J. Pharmacol. Exp. Ther., **325** 1016
- 3) Abian et al. (2011), *Therapeutic strategies for Gaucher disease: miglustat (NB-DNJ) as a pharmacological chaperone for glucocerebrosidase and the different thermostability of velaglycerase alfa and imiglycerase*; Mol. Pharm., **8** 2390
- 4) Serratrice et al. (2015), *Switching from imiglycerase to miglustat for the treatment of French patients with Gaucher disease type 1: a case series*; J. Med. Case Rep., **9** 146
- 5) Karimzadeh (2013), *Effects of miglustat on stabilization of neurological disorder in niemann-pick disease type C: Iranian pediatric case series*; J. Child Neurol., **28** 1599

PHYSICAL DATA

Molecular Weight:	255.74
Molecular Formula:	C ₁₀ H ₂₁ NO ₄ HCl
Purity:	98% by TLC
	NMR: (Conforms)
Solubility:	Soluble in DMSO (up to 20 mg/ml) or in water (up to 20 mg/ml)
Physical Description:	White solid
Storage and Stability:	Store as supplied desiccated at -20°C for up to 1 year from the date of purchase. Solutions in DMSO or distilled water may be stored at -20°C for up to 1 month.

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