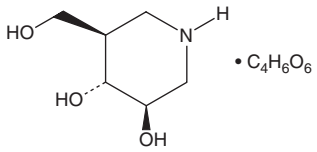
PRODUCT INFORMATION

Isofagomine (D-tartrate)

Item No. 16137

CAS Registry No.: 957230-65-8
Formal Name: 5R-(hydroxymethyl)-3R,4R-piperidinediol, mono 2S,3S-dihydroxybutanedioate
MF: $C_6H_{13}NO_3 \bullet C_4H_6O_6$
FW: 297.3
Purity: ≥95%
Supplied as: A solution in ethanol:water (1:1)
Storage: -20°C
Stability: ≥2 years



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

Laboratory Procedures

Isofagomine (D-tartrate) is supplied as a solution in ethanol:water (1:1). To change the solvent, simply evaporate the ethanol:water (1:1) under a gentle stream of nitrogen and immediately add the solvent of choice. The solvent DMSO purged with an inert gas can be used at a concentration of 2 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. If an organic solvent-free solution of isofagomine (D-tartrate) is needed, it can be prepared by evaporating the ethanol:water and directly dissolving the neat oil in aqueous buffers. The solubility of isofagomine (D-tartrate) in PBS, pH 7.2, is approximately 5 mg/ml. We do not recommend storing the aqueous solution for more than one day

Description

Isofagomine (D-tartrate) is a competitive inhibitor of human lysosomal β-glucosidase (K_i = 0.016-0.025 µM; IC_{50} = 0.06 µM).[1] By interacting with the catalytic pocket of β-glucosidase it acts as a chemical chaperone that increases the amount of β-glucosidase by stabilizing and/or promoting the folding of the enzyme.[2] Isofagomine (D-tartrate) has been shown to increase lysosomal β-glucosidase activity by 2- to 3-fold in mutant N370S Gaucher fibroblasts.[1] This compound has been studied in the context of Gaucher disease, a lysosomal storage disorder resulting from substantial deficiency of β-glucosidase and recently identified as a parkinsonism risk factor.[2]

References

1. Kuriyama, C., Kamiyama, O., Ikeda, K., *et al. In vitro* inhibition of glycogen-degrading enzymes and glycosidases by six-membered sugar mimics and their evaluation in cell cultures. *Bioorg. Med. Chem.* **16(15)**, 7330-7336 (2008).
2. Witte, M.D., Kallemeijn, W.W., Aten, J., *et al*. Ultrasensitive in situ visualization of active glucocerebrosidase molecules. *Nat. Chem. Biol.* **6(12)**, 907-913 (2010).

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