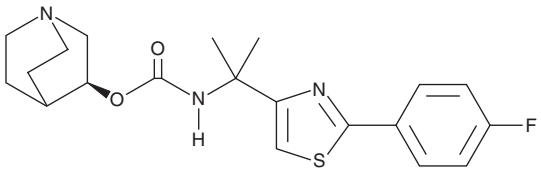
PRODUCT INFORMATION

Ibiglustat

Item No. 32878

CAS Registry No.:	1401090-53-6
Formal Name:	N-[1-[2-(4-fluorophenyl)-4-thiazolyl]-1-methylethyl]-carbamic acid, (3S)-1-azabicyclo[2.2.2]oct-3-yl ester
Synonyms:	Genz-682452, GZ/SAR402671, Venglustat
MF:	$C_{20}H_{24}FN_3O_2S$
FW:	389.5
Purity:	≥98%
UV/Vis.:	λ_{max}: 293 nm
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

Ibiglustat is supplied as a crystalline solid. A stock solution may be made by dissolving the ibiglustat in the solvent of choice, which should be purged with an inert gas. Ibiglustat is soluble in organic solvents such as ethanol, DMSO, and dimethyl formamide (DMF). The solubility of ibiglustat in ethanol and DMF is approximately 30 mg/ml and approximately 25 mg/ml in DMSO.

Ibiglustat is sparingly soluble in aqueous buffers. For maximum solubility in aqueous buffers, ibiglustat should first be dissolved in ethanol and then diluted with the aqueous buffer of choice. Ibiglustat has a solubility of approximately 0.12 mg/ml in a 1:7 solution of ethanol:PBS (pH 7.2) using this method. We do not recommend storing the aqueous solution for more than one day.

Description

Ibiglustat is an inhibitor of glucosylceramide (GlcCer) synthase.[1] Dietary administration of ibiglustat (0.03% w/w) reduces globotriaosylceramide (Gb_3) and lyso-Gb_3 accumulation in a variety of tissues, including the kidney, heart, and brain, in the *Gla*$^{-/-}$ mouse model of Fabry disease. It reduces the latency to paw withdrawal in the hot plate test in the same model, indicating reduced peripheral neuropathy. Ibiglustat (0.03% w/w in the diet) reduces gliosis, delays the onset of ataxia, and increases lifespan in the transgenic 4L;C* mouse model of neuronopathic Gaucher disease.[2]

References

1. Ashe, K.M., Budman, E., Bangari, D.S., *et al.* Efficacy of enzyme and substrate reduction therapy with a novel antagonist of glucosylceramide synthase for Fabry disease. *Mol. Med.* **21(1)**, 389-399 (2015).
2. Marshall, J., Sun, Y., Bangari, D.S., *et al.* CNS-accessible inhibitor of glucosylceramide synthase for substrate reduction therapy of neuronopathic Gaucher disease. *Mol. Ther.* **24(6)**, 1019-1029 (2016).

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