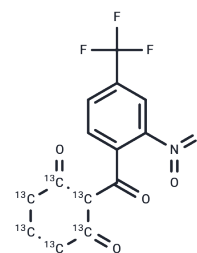


Nitisinone-13C6

Chemical Properties

CAS No. :	1246815-63-3
Formula:	C ₁₄ H ₁₀ F ₃ NO ₅
Molecular Weight:	335.185
Storage:	Powder: -20°C for 3 years In solvent: -80°C for 1 year Actual storage temperature shall be subject to the COA.



Biological Description

Description	<p>Nitisinone-13C6 is intended for use as an internal standard for the quantification of nitisinone by GC- or LC-MS. Nitisinone (T1684) is an inhibitor of 4-hydroxyphenylpyruvate dioxygenase (HPPD), which converts 4-hydroxyphenylpyruvate (HPPA) to homogentisate in the tyrosine catabolic pathway.¹ Nitisinone increases urinary levels of HPPA and 4-hydroxyphenyllactate (HPLA) in rats when administered at a dose of 10 mg/kg. Nitisinone (T1684) (3 mg/kg) prevents the neonatal lethality of fumarylacetoacetate hydrolase (FAH) deficiency in mice when administered to pregnant dams.² It exhibits hepatoprotective effects in FAH^{-/-} mice, such as prevention of increases in plasma levels of aspartate serine aminotransferase (AST) and conjugated bilirubin, when administration is continued following birth at a dose of 1 mg/kg. Nitisinone (T1684) (100 µg) decreases urinary excretion of homogentisate and increases urinary excretion of HPPA, HPLA, and 4-hydroxyphenylacetate in a mouse model of alkaptonuria induced by ethylnitrosourea.³ Formulations containing nitisinone have been used in the treatment of hereditary tyrosinemia type 1 (HT-1).</p>
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Solubility Information

Solubility	<p>Chloroform: Slightly soluble Ethyl Acetate: Slightly soluble (< 1 mg/ml refers to the product slightly soluble or insoluble)</p>
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Preparing Stock Solutions

	1mg	5mg	10mg
1 mM	2.9834 mL	14.9169 mL	29.8338 mL
5 mM	0.5967 mL	2.9834 mL	5.9668 mL
10 mM	0.2983 mL	1.4917 mL	2.9834 mL
50 mM	0.0597 mL	0.2983 mL	0.5967 mL

Please select the appropriate solvent to prepare the stock solution, according to the solubility of the product in different solvents. Please use it as soon as possible.

Note: The dilution table applies only to solid products. For liquid products, please calculate the stock solution based on the stated concentration and/or density.

Reference

Ellis, M.K., Whitfield, A.C., Gowans, L.A., et al. Inhibition of 4-hydroxyphenylpyruvate dioxygenase by 2-(2-nitro-4-trifluoromethylbenzoyl)-cyclohexane-1,3-dione and 2-(2-chloro-4-methanesulfonylbenzoyl)-cyclohexane-1,3-dione. *Toxicol. Appl. Pharmacol.* 133(1)12-19(1995)

Grompe, M., Lindstedt, S., al-Dhalimy, M., et al. Pharmacological correction of neonatal lethal hepatic dysfunction in a murine model of hereditary tyrosinaemia type I. *Nat. Genet.* 10(4)453-460(1995)

Suzuki, Y., Oda, K., Yoshikawa, Y., et al. A novel therapeutic trial of homogentisic aciduria in a murine model of alkaptonuria. *J. Hum. Genet.* 44(2)79-84(1999)

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