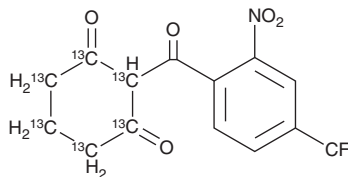


PRODUCT INFORMATION



Nitisinone-¹³C₆ Item No. 28770

CAS Registry No.: 1246815-63-3
Formal Name: 2-(2-nitro-4-(trifluoromethyl)benzoyl)
cyclohexane-1,3-dione-1,2,3,4,5,6-¹³C₆
Synonym: NTBC-¹³C₆
MF: C₈[¹³C₆]H₁₀F₃NO₅
FW: 335.2
Purity: ≥98%
Supplied as: A 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

Nitisinone-¹³C₆ is supplied as a solid. A stock solution may be made by dissolving the nitisinone-¹³C₆ in the solvent of choice, which should be purged with an inert gas. Nitisinone-¹³C₆ is slightly soluble in chloroform and ethyl acetate.

Description

Nitisinone-¹³C₆ is intended for use as an internal standard for the quantification of nitisinone (Item No. 17924) by GC- or LC-MS. Nitisinone 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 (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 (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).

References

1. 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).
2. 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).
3. 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).

WARNING

THIS PRODUCT IS FOR RESEARCH ONLY - NOT FOR HUMAN OR VETERINARY DIAGNOSTIC OR THERAPEUTIC USE.

SAFETY DATA

This material should be considered hazardous until further information becomes available. Do not ingest, inhale, get in eyes, on skin, or on clothing. Wash thoroughly after handling. Before use, the user must review the complete Safety Data Sheet, which has been sent via email to your institution.

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