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Produktinformation

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- Diagnostik & molekulare Diagnostik
- Laborgeräte & Service

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Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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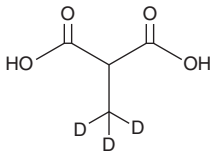
PRODUCT INFORMATION

Methylmalonic Acid-d_3

Item No. 35232

CAS Registry No.: 42522-59-8
Formal Name: 2-methyl-d_3-propanedioic acid
Synonyms: Isosuccinic Acid-d_3, Methylmalonate-d_3, MMA-d_3
MF: $C_4H_3D_3O_4$
FW: 121.1
Chemical Purity: ≥98% (Methylmalonic Acid)
Deuterium Incorporation: ≥99% deuterated forms (d_1-d_3); ≤1% d_0
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

Methylmalonic acid-d_3 is intended for use as an internal standard for the quantification of methylmalonic acid (Item No. 14885) by GC- or LC-MS. The accuracy of the sample weight in this vial is between 5% over and 2% under the amount shown on the vial. If better precision is required, the deuterated standard should be quantitated against a more precisely weighed unlabeled standard by constructing a standard curve of peak intensity ratios (deuterated *versus* unlabeled).

Methylmalonic acid-d_3 is supplied as a solid. A stock solution may be made by dissolving the methylmalonic acid-d_3 in the solvent of choice, which should be purged with an inert gas. Methylmalonic acid-d_3 is slightly soluble in DMSO and methanol.

Description

Methylmalonic acid is a dicarboxylic acid and byproduct of propionate catabolism.[1] It impairs mitochondrial respiration, increases glucose consumption, and downregulates the transcription of the mature neuronal markers neuron-specific enolase (ENO2) and synaptophysin (SYP) in neuronally differentiated SH-SY5Y cells. Intrastriatal administration of methylmalonic acid (100 mg/ml) induces neuronal loss in rats.[2] Methylmalonic acid accumulates in the tissues and body fluids of patients with methylmalonic acidemia, an inborn error of metabolism characterized by a deficiency in the activity of L-methylmaolnyl-CoA mutase or its cofactor 5-deoxyadenosylcobalamin and leads to seizures, intellectual disabilities, psychomotor abnormalities, and coma.[1]

References

1. da Costa, R.T., Dos Santos, M.B., Silva, I.C.S., *et al.* Methylmalonic acid compromises respiration and reduces the expression of differentiation markers of SH-SY5Y human neuroblastoma cells. *ACS Chem. Neurosci.* **12(14)**, 2608-2618 (2021).
2. Narasimhan, P., Sklar, R., Murrell, M., *et al.* Methylmalonyl-CoA mutase induction by cerebral ischemia and neurotoxicity of the mitochondrial toxin methylmalonic acid. *J. Neurosci.* **16(22)**, 7336-7346 (1996).

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