

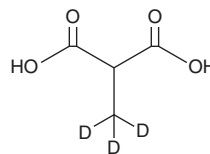
# PRODUCT INFORMATION



## Methylmalonic Acid-d<sub>3</sub>

Item No. 35232

**CAS Registry No.:** 42522-59-8  
**Formal Name:** 2-methyl-d<sub>3</sub>-propanedioic acid  
**Synonyms:** Isosuccinic Acid-d<sub>3</sub>, Methylmalonate-d<sub>3</sub>, MMA-d<sub>3</sub>  
**MF:** C<sub>4</sub>H<sub>3</sub>D<sub>3</sub>O<sub>4</sub>  
**FW:** 121.1  
**Chemical Purity:** ≥98% (Methylmalonic Acid)  
**Deuterium Incorporation:** ≥99% deuterated forms (d<sub>1</sub>-d<sub>3</sub>); ≤1% d<sub>0</sub>  
**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<sub>3</sub> 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<sub>3</sub> is supplied as a solid. A stock solution may be made by dissolving the methylmalonic acid-d<sub>3</sub> in the solvent of choice, which should be purged with an inert gas. Methylmalonic acid-d<sub>3</sub> is slightly soluble in DMSO and methanol.

### Description

Methylmalonic acid is a dicarboxylic acid and byproduct of propionate catabolism.<sup>1</sup> 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. Intrastratial administration of methylmalonic acid (100 mg/ml) induces neuronal loss in rats.<sup>2</sup> 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-methylmalonyl-CoA mutase or its cofactor 5-deoxyadenosylcobalamin and leads to seizures, intellectual disabilities, psychomotor abnormalities, and coma.<sup>1</sup>

### 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).

#### 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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