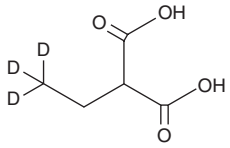
PRODUCT INFORMATION

Ethylmalonic Acid-d_3

Item No. 43923

CAS Registry No.: 70907-93-6
Formal Name: ethyl-2,2,2-d_3-propanedioic acid
Synonym: EMA-d_3
MF: $C_5H_5D_3O_4$
FW: 135.1
Chemical Purity: ≥98% (Ethylmalonic 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

Ethylmalonic acid-d_3 is intended for use as an internal standard for the quantification of ethylmalonic acid 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).

Ethylmalonic acid-d_3 is supplied as a solid. A stock solution may be made by dissolving the ethylmalonic acid-d_3 in the solvent of choice, which should be purged with an inert gas. Ethylmalonic acid-d_3 is soluble (≥10 mg/ml) in ethanol and DMSO.

Description

Ethylmalonic acid is a dicarboxylic acid and metabolite of the essential amino acid L-isoleucine and branched-chain amino acid L-alloisoleucine (Item No. 34904).[1] It is formed from L-isoleucine and L-alloisoleucine *via* an R-2-oxo-3-methylvaleric acid intermediate. Urinary levels of ethylmalonic acid are increased in patients with ethylmalonic encephalopathy, an inborn error of metabolism characterized by developmental delay, hypotonia, vascular instability, petechiae, acrocyanosis, chronic diarrhea, and lactic acidemia. Urinary levels of ethylmalonic acid are also increased in patients with short-chain acyl-CoA dehydrogenase deficiency.[2]

References

1. Nowaczyk, M.J.M., Lehotay, D.C., Platt, B.-A., *et al.* Ethylmalonic and methylsuccinic aciduria in ethylmalonic encephalopathy arise from abnormal isoleucine metabolism. *Metabolism* **47(7),** 836-839 (1998).
2. Gallant, N.M., Leydiker, K., Tang, H., *et al.* Biochemical, molecular, and clinical characteristics of children with short chain acyl-CoA dehydrogenase deficiency detected by newborn screening in California. *Mol. Genet. Metab.* **106(1),** 55-61 (2012).

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