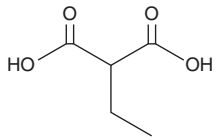
PRODUCT INFORMATION

Ethylmalonic Acid

Item No. 44091

CAS Registry No.: 601-75-2
Formal Name: 2-ethyl-propanedioic acid
MF: $C_5H_8O_4$
FW: 132.1
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

Ethylmalonic acid is supplied as a solid. A stock solution may be made by dissolving the ethylmalonic acid in the solvent of choice, which should be purged with an inert gas. Ethylmalonic acid is sparingly soluble (1-10 mg/ml) in ethanol and slightly soluble (0.1-1 mg/ml) in DMSO.

Further dilutions of the stock solution into aqueous buffers or isotonic saline should be made prior to performing biological experiments. Ensure that the residual amount of organic solvent is insignificant, since organic solvents may have physiological effects at low concentrations. Organic solvent-free aqueous solutions of ethylmalonic acid can be prepared by directly dissolving the solid in aqueous buffers. Ethylmalonic acid is slightly soluble (0.1-1 mg/ml) in PBS (pH 7.2). We do not recommend storing the aqueous solution for more than one day.

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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