

# Produktinformation



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# SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

linkedin.com/company/szaboscandic in



# PRODUCT INFORMATION



## Methylsuccinic Acid

Item No. 36430

CAS Registry No.: 498-21-5

Formal Name: 2-methyl-butanedioic acid Synonyms: 2-Methylsuccinic Acid, NSC 5276

≥4 years

MF:  $C_5H_8O_4$ FW: 132.1 **Purity:** ≥95% Supplied as: A solid Storage: -20°C

Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.

#### **Laboratory Procedures**

Stability:

Methylsuccinic acid is supplied as a solid. A stock solution may be made by dissolving the methylsuccinic acid in the solvent of choice, which should be purged with an inert gas. Methylsuccinic acid is soluble in organic solvents such as ethanol, DMSO, and dimethyl formamide. The solubility of methylsuccinic acid in these solvents is approximately 30 mg/ml.

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 methylsuccinic acid can be prepared by directly dissolving the solid in aqueous buffers. The solubility of methylsuccinic acid in PBS (pH 7.2) is approximately 10 mg/ml. We do not recommend storing the aqueous solution for more than one day.

### Description

Methylsuccinic acid is a dicarboxylic acid and metabolite of the essential amino acid L-isoleucine and branched-chain amino acid L-alloisoleucine (Item No. 34904).<sup>1</sup> It is formed from L-isoleucine and L-alloisoleucine via an R-2-oxo-3-methylvaleric acid intermediate. Urinary levels of methylsuccinic 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 methylsuccinic acid are also increased in patients with short-chain acyl-CoA dehydrogenase deficiency or type 2 diabetes.<sup>2,3</sup> It has been found in urban and industrial aerosols of fine particulate matter less than 2.5 μm (PM<sub>2.5</sub>).<sup>4</sup>

#### 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
- 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).
- 3. Urpi-Sarda, M., Almanza-Aguilera, E., Llorach, R., et al. Non-targeted metabolomic biomarkers and metabotypes of type 2 diabetes: A cross-sectional study of PREDIMED trial participants. Diabetes Metab. **45(2)**, 167-174 (2019).
- 4. Crenn, V., Fronval, I., Petitprez, D., et al. Fine particles sampled at an urban background site and an industrialized coastal site in Northern France - Part 1: Seasonal variations and chemical characterization. Sci. Total Environ. 578, 203-218 (2017).

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

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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## **CAYMAN CHEMICAL**

1180 EAST ELLSWORTH RD ANN ARBOR, MI 48108 · USA PHONE: [800] 364-9897

[734] 971-3335

FAX: [734] 971-3640 CUSTSERV@CAYMANCHEM.COM WWW.**CAYMANCHEM**.COM