

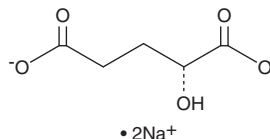
PRODUCT INFORMATION



D- α -Hydroxyglutaric Acid (sodium salt)

Item No. 11605

CAS Registry No.: 103404-90-6
Formal Name: 2R-hydroxy-pentanedioic acid, disodium salt
Synonyms: D-2-HG, D-2-Hydroxyglutaric Acid
MF: C₅H₆O₅ • 2Na
FW: 192.1
Purity: ≥95%
Stability: ≥2 years at -20°C
Supplied as: A crystalline solid



Laboratory Procedures

For long term storage, we suggest that D- α -hydroxyglutaric acid (D-2-HG) (sodium salt) be stored as supplied at -20°C. It should be stable for at least two years.

D-2-HG (sodium salt) is supplied as a crystalline solid. D-2-HG (sodium salt) is sparingly soluble in organic solvents such as ethanol, DMSO, and dimethyl formamide. For biological experiments, we suggest that organic solvent-free aqueous solutions of D-2-HG (sodium salt) be prepared by directly dissolving the crystalline solid in aqueous buffers. The solubility of D-2-HG (sodium salt) in PBS, pH 7.2, is approximately 10 mg/ml. We do not recommend storing the aqueous solution for more than one day.

Description

D-2-HG is an α -hydroxy acid that is over-produced in the human neurometabolic disease D-2-hydroxyglutaric aciduria (D-2-HGA).¹ It is normally synthesized from 2-ketoglutarate (2-KG) by hydroxyacid-oxoacid transhydrogenase (HOT), although defects in HOT are not known to be associated with D-2-HGA.¹ Instead, type I D-2-HGA involves mutations in D-2-hydroxyglutarate dehydrogenase, which converts D-2-HG back to 2-KG.¹ Type II D-2-HGA results from gain-of-function mutations in isocitrate dehydrogenase 2, causing it to supplement HOT in converting 2-KG to D-2-HG.^{2,3} In bacteria, this α -hydroxy acid may be synthesized from oxalate- and propionyl-coenzyme A by an α -hydroxyglutaric acid synthetase.⁴

References

1. Kranendijk, M., Struys, E.A., Salomons, G.S., *et al.* Progress in understanding 2-hydroxyglutaric acidurias. *J. Inherit. Metab. Dis.* **35(4)**, 571-587 (2012).
2. Kranendijk, M., Struys, E.A., Van Schaftingen, E., *et al.* IDH2 Mutations in Patients with D-2-Hydroxyglutaric Aciduria. *Science* **330(6002)**, 336 (2014).
3. Struys, E.A., Salomons, G.S., Achouri, Y., *et al.* Mutations in the D-2-hydroxyglutarate dehydrogenase gene cause D-2-hydroxyglutaric aciduria. *Am. J. Hum. Genet.* **76**, 358-360 (2005).
4. Reeves, H.C. and Ajl, S.J. Alpha-hydroxyglutaric acid synthetase. *J. Bacteriol.* **84**, 186-187 (1962).

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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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