

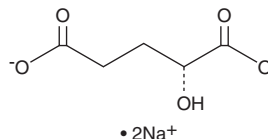
PRODUCT INFORMATION



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%
Supplied as: A crystalline 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

D- α -hydroxyglutaric acid (D-2-HG) (sodium salt) is supplied as a crystalline solid. Aqueous solutions of D-2-HG (sodium salt) can 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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