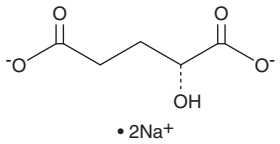
PRODUCT INFORMATION

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: $C_5H_6O_5 \bullet 2Na$
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).

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