

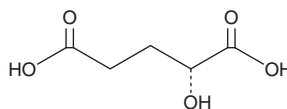
# PRODUCT INFORMATION



## D- $\alpha$ -Hydroxyglutaric Acid

Item No. 25895

CAS Registry No.: 13095-47-1  
Formal Name: 2R-hydroxy-pentanedioic acid  
Synonyms: D-2-HG, D-2-Hydroxyglutaric Acid  
MF:  $C_5H_8O_5$   
FW: 148.1  
Purity:  $\geq 95\%$   
UV/Vis.:  $\lambda_{max}$ : 212 nm  
Supplied as: A solution in ethanol  
Storage:  $-20^\circ C$   
Stability:  $\geq 1$  year



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

### Laboratory Procedures

D- $\alpha$ -Hydroxyglutaric acid (D-2-HG) is supplied as a solution in ethanol. To change the solvent, simply evaporate the ethanol under a gentle stream of nitrogen and immediately add the solvent of choice. Solvents such as DMSO and dimethyl formamide purged with an inert gas can be used. The solubility of D-2-HG 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. If an organic solvent-free solution of D-2-HG is needed, it can be prepared by evaporating the ethanol and directly dissolving the neat oil in aqueous buffers. The solubility of D-2-HG 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  $\alpha$ -hydroxy acid that is over-produced in the human neurometabolic disease D-2-hydroxyglutaric aciduria (D-2-HGA).<sup>1</sup> 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.<sup>1</sup> Instead, type I D-2-HGA involves mutations in D-2-hydroxyglutarate dehydrogenase, which converts D-2-HG back to 2-KG.<sup>1</sup> Type II D-2-HGA results from gain-of-function mutations in isocitrate dehydrogenase 2 (IDH2), causing it to supplement HOT in converting 2-KG to D-2-HG.<sup>2,3</sup> In bacteria, this  $\alpha$ -hydroxy acid may be synthesized from oxalate and propionyl-coenzyme A by an  $\alpha$ -hydroxyglutaric acid synthetase.<sup>4</sup>

### 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**(2), 358-360 (2005).
4. Reeves, H.C. and Ajl, S.J. Alpha-hydroxyglutaric acid synthetase. *J. Bacteriol.* **84**(1), 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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