

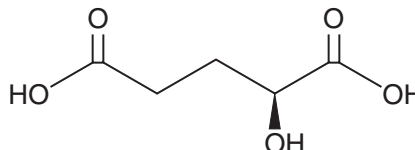
# PRODUCT INFORMATION



## L- $\alpha$ -Hydroxyglutaric Acid

Item No. 21123

**CAS Registry No.:** 13095-48-2  
**Formal Name:** 2S-hydroxy-pentanedioic acid  
**Synonyms:** 2(S)-HG, 2(S)-Hydroxyglutaric Acid, L-2-HG, L-2-Hydroxyglutaric Acid,  
**MF:** C<sub>5</sub>H<sub>8</sub>O<sub>5</sub>  
**FW:** 148.1  
**Purity:**  $\geq$ 95%  
**Supplied as:** A solution in ethanol  
**Storage:** -20°C  
**Stability:**  $\geq$ 1 year



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

### Laboratory Procedures

L- $\alpha$ -Hydroxyglutaric acid is supplied as a solution in ethanol. To change the solvent, simply evaporate the L- $\alpha$ -hydroxyglutaric acid under a gentle stream of nitrogen and immediately add the solvent of choice. Solvents such as ethanol, DMSO, and dimethyl formamide purged with an inert gas can be used. The solubility of L- $\alpha$ -hydroxyglutaric 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. If an organic solvent-free solution of L- $\alpha$ -hydroxyglutaric acid is needed, it can be prepared by evaporating the ethanol and directly dissolving the neat oil in aqueous buffers. The solubility of L- $\alpha$ -hydroxyglutaric 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

L- $\alpha$ -Hydroxyglutaric acid (L-2-HG) is an  $\alpha$ -hydroxy acid. It is metabolized to 2-oxoglutarate ( $\alpha$ -ketoglutarate) by L-2-hydroxyglutarate dehydrogenase, and mutations in this enzyme lead to 2-hydroxyglutaric aciduria, a neurometabolic disorder characterized by increased L-2-HG levels.<sup>1,2</sup> L-2-HG is structurally similar to  $\alpha$ -ketoglutarate and competitively inhibits  $\alpha$ -ketoglutarate-dependent dioxygenases, including several involved in histone lysine and DNA demethylation.<sup>3,4</sup>

### References

1. Rzem, R., Veiga-da-Cunha, M., Noël, G., *et al.* A gene encoding a putative FAD-dependent L-2-hydroxyglutarate dehydrogenase is mutated in L-2-hydroxyglutaric aciduria. *Proc. Nat. Acad. Sci. USA* **101(48)**, 16849-16854 (2004).
2. Struys, E.A., Verhoeven, N.M., Roos, B., *et al.* Disease-related metabolites in culture medium of fibroblasts from patients with D-2-hydroxyglutaric aciduria, L-2-hydroxyglutaric aciduria, and combined D/L-2-hydroxyglutaric aciduria. *Clin. Chem.* **49(7)**, 1133-1138 (2003).
3. Chowdhury, R., Yeoh, K.K., Tian, Y.M., *et al.* The oncometabolite 2-hydroxyglutarate inhibits histone lysine demethylases. *EMBO Rep.* **12(5)**, 463-469 (2011).
4. Xu, W., Yang, H., Liu, Y., *et al.* Oncometabolite 2-hydroxyglutarate is a competitive inhibitor of  $\alpha$ -ketoglutarate-dependent dioxygenases. *Cancer Cell* **19(1)**, 17-30 (2011).

#### 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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