

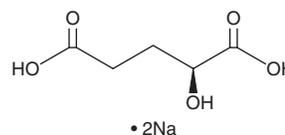
# PRODUCT INFORMATION



## L- $\alpha$ -Hydroxyglutaric Acid (sodium salt)

Item No. 11876

**CAS Registry No.:** 63512-50-5  
**Formal Name:** 2S-hydroxy-pentanedioic acid, disodium salt  
**Synonyms:** L-2-HG, L-2-Hydroxyglutarate, L-2-Hydroxyglutaric Acid, (S)-2-Hydroxypentanedioic Acid  
**MF:** C<sub>5</sub>H<sub>8</sub>O<sub>5</sub> • 2Na  
**FW:** 194.1  
**Purity:** ≥95%  
**Supplied as:** A crystalline solid  
**Storage:** -20°C  
**Stability:** ≥2 years



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

### Laboratory Procedures

L- $\alpha$ -Hydroxyglutaric acid (sodium salt) is supplied as a crystalline solid. Aqueous solutions of L- $\alpha$ -hydroxyglutaric acid (sodium salt) can be prepared by directly dissolving the crystalline solid in aqueous buffers. The solubility of L- $\alpha$ -hydroxyglutaric acid (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

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.

#### WARRANTY AND LIMITATION OF REMEDY

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