

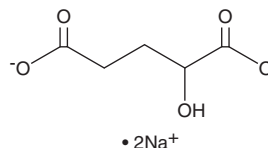
# PRODUCT INFORMATION



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

Item No. 16374

**CAS Registry No.:** 40951-21-1  
**Formal Name:** 2-hydroxy-pentanedioic acid, disodium salt  
**Synonyms:** FA 5:1;O3, 2-HG, 2-Hydroxyglutaric Acid  
**MF:** C<sub>5</sub>H<sub>6</sub>O<sub>5</sub> • 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

$\alpha$ -Hydroxyglutaric acid (2-HG) (sodium salt) is supplied as a crystalline solid. Aqueous solutions of 2-HG (sodium salt) can be prepared by directly dissolving the crystalline solid in aqueous buffers. The solubility of 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

2-HG is an  $\alpha$ -hydroxy acid. It is metabolized to 2-oxoglutarate by D- and L-2-hydroxyglutarate dehydrogenases, and mutations in these enzymes lead to 2-hydroxyglutaric aciduria, a neurometabolic disorder characterized by increased levels of L-2-hydroxyglutaric acid (Item No. 21123).<sup>1-3</sup> 2-HG can also be metabolized from isocitrate by mutated forms of isocitrate dehydrogenase 1 (IDH1) and IDH2.<sup>4,5</sup> 2-HG is structurally similar to  $\alpha$ -ketoglutarate, the product of wild-type isocitrate dehydrogenases, and competitively inhibits  $\alpha$ -ketoglutarate-dependent dioxygenases, including histone lysine demethylases and DNA hydroxylases.<sup>5-7</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. Natl. 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. 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. Ward, P., Patel, J., Wise, D.R., *et al.* The common feature of leukemia-associated IDH1 and IDH2 mutations is a neomorphic enzyme activity converting  $\alpha$ -ketoglutarate to 2-hydroxyglutarate. *Cancer Cell* **17(3)**, 225-234 (2010).
5. Yang, H., Ye, D., Guan, K.-L., *et al.* IDH1 and IDH2 mutations in tumorigenesis: Mechanistic insights and clinical perspectives. *Clin. Cancer Res.* **18(20)**, 5562-5571 (2012).
6. 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).
7. 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).

#### 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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