# HGD (H-133): sc-98503



The Power to Question

### **BACKGROUND**

HGD (homogentisate 1,2-dioxygenase), also known as HGO (homogentisate oxygenase), is a 445 amino acid protein that belongs to the homogentisate dioxygenase family and is involved in the pathway of amino acid degradation. Expressed at high levels in kidney, colon, liver, prostate and small intestine, HGD uses iron as a cofactor to catalyze the oxygen-dependent conversion of homogentisate to 4-maleylacetoacetate, a reaction that is the fourth step in the creation of L-phenylalanine from fumarate and acetoacetic acid. Defects in the gene encoding HGD are the cause of alkaptonuria (AKU), an autosomal recessive disorder that is characterized by urine that turns dark on standing and alkalinization, black ochronotic pigmentation of cartilage and collagenous tissues and spine arthritis.

## **REFERENCES**

- Pollak, M.R., et al. 1993. Homozygosity mapping of the gene for alkaptonuria to chromosome 3q2. Nat. Genet. 5: 201-204.
- 2. Janocha, S., et al. 1994. The human gene for alkaptonuria (AKU) maps to chromosome 3q. Genomics 19: 5-8.
- 3. Hudecová, S., et al. 1995. Purification of the homogentisic acid oxidase from mammalian liver. Int. J. Biochem. Cell Biol. 27: 1357-1363.
- 4. Granadino, B., et al. 1997. The human homogentisate 1,2-dioxygenase (HGO) gene. Genomics 43: 115-122.
- Beltrán-Valero de Bernabe, D., et al. 1998. Mutation and polymorphism analysis of the human homogentisate 1, 2-dioxygenase gene in alkaptonuria patients. Am. J. Hum. Genet. 62: 776-784.
- Ramos, S.M., et al. 1998. Molecular diagnosis of alkaptonuria mutation by analysis of homogentisate 1,2 dioxygenase mRNA from urine and blood. Am. J. Med. Genet. 78: 192-194.
- Beltrán-Valero de Bernabe, D., et al. 1999. Analysis of alkaptonuria (AKU) mutations and polymorphisms reveals that the CCC sequence motif is a mutational hot spot in the homogentisate 1,2 dioxygenase gene (HGO). Am. J. Hum. Genet. 64: 1316-1322.
- 8. Titus, G.P., et al. 2000. Crystal structure of human homogentisate dioxygenase. Nat. Struct. Biol. 7: 542-546.
- 9. Abdulrazzaq, Y.M., et al. 2009. R58fs mutation in the HGD gene in a family with alkaptonuria in the UAE. Ann. Hum. Genet. 73: 125-130.

## **CHROMOSOMAL LOCATION**

Genetic locus: HGD (human) mapping to 3q13.33; Hgd (mouse) mapping to 16 B3.

## SOURCE

HGD (H-133) is a rabbit polyclonal antibody raised against amino acids 97-229 mapping near the N-terminus of HGD of human origin.

#### **PRODUCT**

Each vial contains 200  $\mu g$  lgG in 1.0 ml of PBS with <0.1% sodium azide and 0.1% gelatin.

#### **APPLICATIONS**

HGD (H-133) is recommended for detection of HGD of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

HGD (H-133) is also recommended for detection of HGD in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for HGD siRNA (h): sc-75249, HGD siRNA (m): sc-75250, HGD shRNA Plasmid (h): sc-75249-SH, HGD shRNA Plasmid (m): sc-75250-SH, HGD shRNA (h) Lentiviral Particles: sc-75249-V and HGD shRNA (m) Lentiviral Particles: sc-75250-V.

Molecular Weight of HGD: 50 kDa.

Positive Controls: Mouse liver extract: sc-2256.

#### **RECOMMENDED SECONDARY REAGENTS**

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

#### **STORAGE**

Store at  $4^{\circ}$  C, \*\*DO NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

# **RESEARCH USE**

For research use only, not for use in diagnostic procedures.

#### **PROTOCOLS**

See our web site at www.scbt.com or our catalog for detailed protocols and support products.

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