

HIBADH (D-15): sc-243020

BACKGROUND

HIBADH (3-hydroxyisobutyrate dehydrogenase) is a 336 amino acid mitochondrial enzyme that catalyzes the NAD⁺-dependent, reversible oxidization of 3-Hydroxyisobutyrate to methylmalonate semialdehyde, an intermediate of valine catabolism. The enzyme functions as a homodimer between a pH of 7.0 and 10.0, with optimal activity between 8.8 and 9.0. It was previously hypothesized that defects in the gene encoding HIBADH may be the cause of 3-hydroxyisobutyric aciduria, a rare disorder that is characterized by a variety of clinical manifestations such as neurodevelopmental problems and dysmorphic features. However, it was shown that HIBADH activity was equal in patients with 3-hydroxyisobutyric aciduria as compared with controls.

REFERENCES

1. Rougraff, P.M., et al. 1989. Cloning and sequence analysis of a cDNA for 3-hydroxyisobutyrate dehydrogenase. Evidence for its evolutionary relationship to other pyridine nucleotide-dependent dehydrogenases. *J. Biol. Chem.* 264: 5899-5903.
2. Lokanath, N.K., et al. 2003. Crystallization and preliminary X-ray crystallographic studies of NADP-dependent 3-hydroxyisobutyrate dehydrogenase from *Thermus thermophilus* HB8. *Acta Crystallogr. D Biol. Crystallogr.* 59: 2294-2296.
3. Lehoczy, J.A., et al. 2004. Conserved expression domains for genes upstream and within the HoxA and HoxD clusters suggests a long-range enhancer existed before cluster duplication. *Evol. Dev.* 6: 423-430.
4. Online Mendelian Inheritance in Man, OMIM™. 2004. Johns Hopkins University, Baltimore, MD. MIM Number: 608475. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
5. Lokanath, N.K., et al. 2005. Crystal structure of novel NADP-dependent 3-hydroxyisobutyrate dehydrogenase from *Thermus thermophilus* HB8. *J. Mol. Biol.* 352: 905-917.

CHROMOSOMAL LOCATION

Genetic locus: HIBADH (human) mapping to 7p15.2; Hibadh (mouse) mapping to 6 B3.

SOURCE

HIBADH (D-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of HIBADH of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-243020 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

HIBADH (D-15) is recommended for detection of HIBADH of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg 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).

HIBADH (D-15) is also recommended for detection of HIBADH in additional species, including equine, canine and bovine.

Suitable for use as control antibody for HIBADH siRNA (h): sc-89756, HIBADH siRNA (m): sc-145957, HIBADH shRNA Plasmid (h): sc-89756-SH, HIBADH shRNA Plasmid (m): sc-145957-SH, HIBADH shRNA (h) Lentiviral Particles: sc-89756-V and HIBADH shRNA (m) Lentiviral Particles: sc-145957-V.

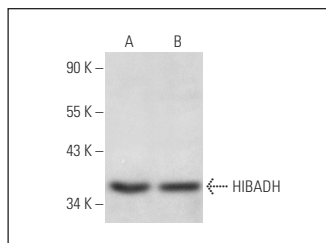
Molecular Weight of HIBADH: 35 kDa.

Positive Controls: CCD-1064Sk cell lysate: sc-2263 or BJ whole cell lysate: sc-364359.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



HIBADH (D-15): sc-243020. Western blot analysis of HIBADH expression in CCD-1064Sk (A) and BJ (B) whole cell lysates.

RESEARCH USE

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



Try **HIBADH (D-11): sc-398288**, our highly recommended monoclonal alternative to HIBADH (D-15).