BACKGROUND

HIBADH (3-hydroxyisobutyrate dehydrogenase) is a 336 amino acid mitochondrial enzyme that catalyzes the NAD+-dependent, reversible oxidation 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


CHROMOSOMAL LOCATION

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

SOURCE

HIBADH (D-11) is a mouse monoclonal antibody raised against amino acids 1-157 mapping at the N-terminus of HIBADH of human origin.

PRODUCT

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

APPLICATIONS

HIBADH (D-11) is recommended for detection of HIBADH of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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).


Molecular Weight of HIBADH: 35 kDa.

Positive Controls: A549 cell lysate: sc-2413, Hep G2 cell lysate: sc-2227 or CCD-1064Sk cell lysate: sc-2263.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), 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 m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Hard-set Mounting Medium: sc-24941 or UltraCruz® Mounting Medium: sc-359850.

DATA

Store at 4° 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 for detailed protocols and support products.