SANTA CRUZ BIOTECHNOLOGY, INC.

D2HGDH (E-6): sc-514171

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

D2HGDH (D-2-hydroxyglutarate dehydrogenase), also known as FLJ42195 or MGC25181, is a 521 amino acid member of the FAD-binding oxidoreductase/transferase type 4 protein family. Localized to mitochondria, D2HGDH is activated by cobalt and zinc and utilizes FAD as a cofactor. D2HGDH catalyzes the oxidation of D-2-hydroxyglutarate, resulting in α-ketoglutarate. Defects in the gene that encodes D2HGDH are the cause of D-2-hydroxyglutaric aciduria (D2HGA), a rare recessive neurometabolic disorder characterized by early infantile-onset epileptic encephalopathy and cardiomyopathy. D2HGA causes developmental delay, hypotonia, epilepsy and dysmorphic features. D2HGDH contains one FAD-binding PCMH-type domain and is expressed as two isoforms produced by alternative splicing.

REFERENCES

1. Gibson, K.M., et al. 1993. D-2-hydroxyglutaric aciduria in a newborn with neurological abnormalities: a new neurometabolic disorder? J. Inherit. Metab. Dis. 16: 497-500.
2. Achouri, Y., et al. 2004. Identification of a dehydrogenase acting on D-2-hydroxyglutarate. Biochem. J. 381: 35-42.
3. Struys, E.A., et al. 2005. Mutations in the D-2-hydroxyglutarate dehydrogenase gene cause D-2-hydroxyglutaric aciduria. Am. J. Hum. Genet. 76: 358-360.
4. Struys, E.A., et al. 2005. Mutations in phenotypically mild D-2-hydroxyglutaric aciduria. Ann. Neurol. 58: 626-630.
5. Misra, V.K., et al. 2005. Phenotypic heterogeneity in the presentation of D-2-hydroxyglutaric aciduria in monozygotic twins. Mol. Genet. Metab. 86: 200-205.
6. Struys, E.A., et al. 2006. D-2-hydroxyglutaric aciduria in three patients with proven SSADH deficiency: genetic coincidence or a related biochemical epiphenomenon? Mol. Genet. Metab. 88: 53-57.

CHROMOSOMAL LOCATION

Genetic locus: D2HGDH (human) mapping to 2q37.3.

SOURCE

D2HGDH (E-6) is a mouse monoclonal antibody raised against amino acids 41-146 mapping near the N-terminus of D2HGDH of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

D2HGDH (E-6) is available conjugated to agarose (sc-514171 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-514171 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514171 PE), fluorescein (sc-514171 FITC), Alexa Fluor® 488 (sc-514171 AF488), Alexa Fluor® 546 (sc-514171 AF546), Alexa Fluor® 594 (sc-514171 AF594) or Alexa Fluor® 647 (sc-514171 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514171 AF680) or Alexa Fluor® 790 (sc-514171 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

D2HGDH (E-6) is recommended for detection of D2HGDH of 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 (predicted) of D2HGDH isoforms: 56/33 kDa.

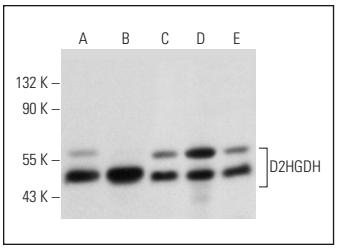
Molecular Weight (observed) of D2HGDH isoforms: 50 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, PANC-1 whole cell lysate: sc-364380 or Jurkat whole cell lysate: sc-2204.

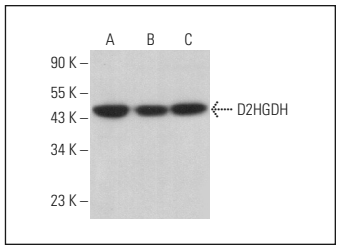
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



D2HGDH (E-6): sc-514171. Western blot analysis of D2HGDH expression in PANC-1 (**A**), Hep G2 (**B**), HeLa (**C**), Jurkat (**D**) and JAR (**E**) whole cell lysates.



D2HGDH (E-6): sc-514171. Western blot analysis of D2HGDH expression in Hep G2 (**A**), K-562 (**B**) and HL-60 (**C**) whole cell lysates.

STORAGE

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.