SANTA CRUZ BIOTECHNOLOGY, INC.

IVD (A-8): sc-514240

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

IVD (isovaleryl-CoA dehydrogenase, mitochondrial) is a 423 amino acid protein encoded by the human gene IVD. IVD is a mitochondrion matrix protein that belongs to the acyl-CoA dehydrogenase family. IVD is a homotetrameric flavoenzyme which catalyzes the conversion of isovaleryl-CoA to 3-methylcrotonyl-CoA. Defects of the IVD gene lead to ineffective isoforms that are the underlying cause of isovaleric acidemia. Two forms of isovaleric acidemia, possibly allelic, are recognized: the acute neonatal form, leading to massive metabolic acidosis from the first days of life and rapid death, and a chronic form in which periodic attacks of severe ketoacidosis occur with asymptomatic intervening periods. There are seven classes of mutants, each with different deletions and pathologies.

REFERENCES

1. Vockley, J., et al. 1992. The variant human isovaleryl-CoA dehydrogenase gene responsible for type II isovaleric acidemia determines an RNA splicing error, leading to the deletion of the entire second coding exon and the production of a truncated precursor protein that interacts poorly with mitochondrial import receptors. J. Biol. Chem. 267: 2494-2501.
2. Parimoo, B. and Tanaka, K. 1993. Structural organization of the human isovaleryl-CoA dehydrogenase gene. Genomics 15: 582-590.
3. Vockley, J., et al. 2000. Exon skipping in IVD RNA processing in isovaleric acidemia caused by point mutations in the coding region of the IVD gene. Am. J. Hum. Genet. 66: 356-367.
4. Tajima, G., et al. 2005. Establishment of a practical enzymatic assay method for determination of isovaleryl-CoA dehydrogenase activity using high-performance liquid chromatography. Clin. Chim. Acta 353: 193-199.
5. Goetzman, E.S., et al. 2006. Functional analysis of acyl-CoA dehydrogenase catalytic residue mutants using surface plasmon resonance and circular dichroism. Mol. Genet. Metab. 87: 233-242.

CHROMOSOMAL LOCATION

Genetic locus: IVD (human) mapping to 15q15.1; Ivd (mouse) mapping to 2 E5.

SOURCE

IVD (A-8) is a mouse monoclonal antibody raised against amino acids 274-396 mapping near the C-terminus of IVD of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

IVD (A-8) is available conjugated to agarose (sc-514240 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-514240 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514240 PE), fluorescein (sc-514240 FITC), Alexa Fluor® 488 (sc-514240 AF488), Alexa Fluor® 546 (sc-514240 AF546), Alexa Fluor® 594 (sc-514240 AF594) or Alexa Fluor® 647 (sc-514240 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514240 AF680) or Alexa Fluor® 790 (sc-514240 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

IVD (A-8) is recommended for detection of IVD, mitochondrial isoform 1 and 2 precursor 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).

Suitable for use as control antibody for IVD siRNA (h): sc-62511, IVD siRNA (m): sc-62512, IVD shRNA Plasmid (h): sc-62511-SH, IVD shRNA Plasmid (m): sc-62512-SH, IVD shRNA (h) Lentiviral Particles: sc-62511-V and IVD shRNA (m) Lentiviral Particles: sc-62512-V.

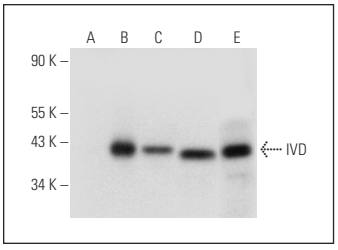
Molecular Weight of IVD: 45 kDa.

Positive Controls: IVD (m): 293T Lysate: sc-127026, HeLa whole cell lysate: sc-2200 or human cervix extract: sc-363756.

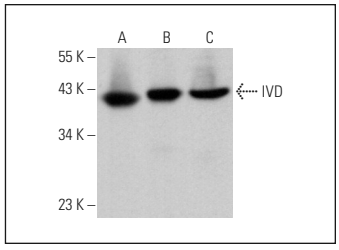
RECOMMENDED SUPPORT REAGENTS

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



IVD (A-8): sc-514240. Western blot analysis of IVD expression in non-transfected 293T: sc-117752 (**A**), mouse IVD transfected 293T: sc-127026 (**B**), F9 (**C**) and HeLa (**D**) whole cell lysates and human cervix tissue extract (**E**).



IVD (A-8): sc-514240. Western blot analysis of IVD expression in human liver (**A**), mouse kidney (**B**) and rat heart (**C**) tissue extracts.

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.