

IVD (B-9): sc-271205

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

- 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.
- Parimoo, B. and Tanaka, K. 1993. Structural organization of the human isovaleryl-CoA dehydrogenase gene. *Genomics* 15: 582-590.
- 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.
- 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.
- 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.
- Vockley, J. and Ensenauer, R. 2006. Isovaleric acidemia: new aspects of genetic and phenotypic heterogeneity. *Am. J. Med. Genet. C, Semin. Med. Genet.* 142: 95-103.

CHROMOSOMAL LOCATION

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

SOURCE

IVD (B-9) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 143-186 within an internal region of IVD of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-271205 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

IVD (B-9) is recommended for detection of IVD 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

IVD (B-9) is also recommended for detection of IVD in additional species, including canine and bovine.

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.

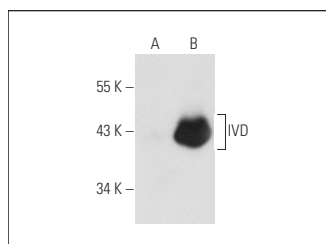
Molecular Weight of IVD: 45 kDa.

Positive Controls: IVD (m): 293T Lysate: sc-127026.

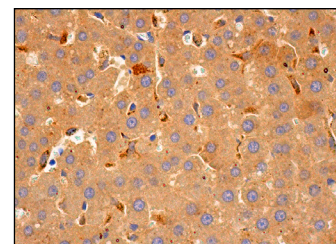
RECOMMENDED SUPPORT REAGENTS

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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



IVD (B-9): sc-271205. Western blot analysis of IVD expression in non-transfected: sc-117752 (A) and mouse IVD transfected: sc-127026 (B) 293T whole cell lysates.



IVD (B-9): sc-271205. Immunoperoxidase staining of formalin fixed, paraffin-embedded human liver tissue showing cytoplasmic staining of hepatocytes.

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.