SANTA CRUZ BIOTECHNOLOGY, INC.

IVD (G-17): sc-55720

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-methyl-crotonyl-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., Nagao, M., Parimoo, B. and Tanaka, K. 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., Rogan, P.K., Anderson, B.D., Willard, J., Seelan, R.S., Smith, D.I. and Liu, W. 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., Sakura, N., Yofune, H., Dwi Bahagia Febriani, A., Nishimura, Y., Sakamoto, A., Ono, H., Shigematsu, Y. and Kobayashi, M. 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.

CHROMOSOMAL LOCATION

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

SOURCE

IVD (G-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of IVD 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-55720 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

IVD (G-17) is recommended for detection of Isovaleryl-CoA dehydrogenase 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).

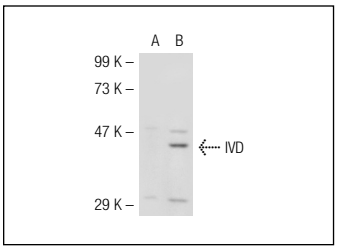
IVD (G-17) is also recommended for detection of Isovaleryl-CoA dehydrogenase in additional species, including equine, canine, bovine, porcine and avian.

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.

DATA



IVD (G-17): sc-55720. Western blot analysis of IVD expression in non-transfected: sc-117752 (**A**) and mouse IVD transfected: sc-127026 (**B**) 293T whole cell lysates.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.