

IVD (m): 293T Lysate: sc-127026

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

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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.
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5. Goetzman, E.S., He, M., Nguyen, T.V. and Vockley, J. 2006. Functional analysis of acyl-CoA dehydrogenase catalytic residue mutants using surface plasmon resonance and circular dichroism. *Mol. Genet. Metab.* 87: 233-242.
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STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

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

CHROMOSOMAL LOCATION

Genetic locus: Ivd (mouse) mapping to 2 E5.

PRODUCT

IVD (m): 293T Lysate represents a lysate of mouse IVD transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

IVD (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive IVD antibodies. Recommended use: 10-20 µl per lane.

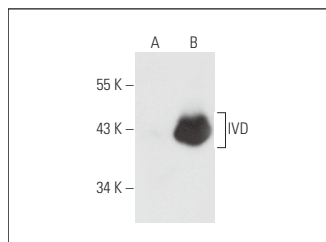
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

IVD (B-9): sc-271205 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse IVD expression in IVD transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

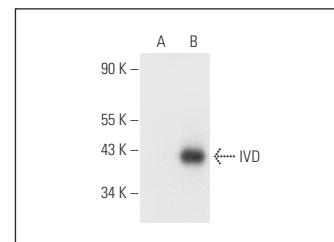
RECOMMENDED SUPPORT REAGENTS

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.

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 (A-8): sc-514240. 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.