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Lieferung & Zahlungsart

siehe unsere Liefer- und Versandbedingungen

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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SANTA CRUZ BIOTECHNOLOGY, INC.

HPPD (h): 293 Lysate: sc-159724

BACKGROUND

HPPD (4-hydroxyphenylpyruvate dioxygenase), also known as PPD, GLOD3 or HPD, is a 393 amino acid protein that belongs to the 4HPPD family and is involved in amino acid degradation. Existing as a homodimer, HPPD uses zinc as a cofactor to catalyze the third step in the conversion of L-phenylalanine to fumarate and acetoacetic acid. Defects in the gene encoding HPPD are the cause of tyrosinemia type 3 (TYRO3) and hawkinsinuria (HAWK), both of which are inborn errors of metabolism that are associated with a variety of symptoms, including mental retardation and seizures (associated with TYRO3) and hair and urine abnormalities (associated with HAWK). The gene encoding HPPD maps to human chromosome 12, which encodes over 1,100 genes and comprises approximately 4.5% of the human genome.

REFERENCES

1. Rüetschi, U., Dellsén, A., Sahlin, P., Stenman, G., Rymo, L. and Lindstedt, S. 1993. Human 4-hydroxyphenylpyruvate dioxygenase. Primary structure and chromosomal localization of the gene. Eur. J. Biochem. 213: 1081-1089.
2. Awata, H., Endo, F. and Matsuda, I. 1994. Structure of the human 4-hydroxyphenylpyruvic acid dioxygenase gene (HPD). Genomics 23: 534-539.
3. Stenman, G., Röijer, E., Rüetschi, U., Dellsén, A., Rymo, L. and Lindstedt, S. 1995. Regional assignment of the human 4-hydroxyphenylpyruvate dioxygenase gene (HPD) to 12q24→qter by fluorescence *in situ* hybridization. Cytogenet. Cell Genet. 71: 374-376.
4. Rüetschi, U., Rymo, L. and Lindstedt, S. 1997. Human 4-hydroxyphenylpyruvate dioxygenase gene (HPD). Genomics 44: 292-299.
5. Rüetschi, U., Cerone, R., Pérez-Cerda, C., Schiaffino, M.C., Standing, S., Ugarte, M. and Holme, E. 2000. Mutations in the 4-hydroxyphenylpyruvate dioxygenase gene (HPD) in patients with tyrosinemia type III. Hum. Genet. 106: 654-662.
6. Tomoeda, K., Awata, H., Matsuura, T., Matsuda, I., Ploechl, E., Milovac, T., Boneh, A., Scott, C.R., Danks, D.M. and Endo, F. 2000. Mutations in the 4-hydroxyphenylpyruvic acid dioxygenase gene are responsible for tyrosinemia type III and hawkinsinuria. Mol. Genet. Metab. 71: 506-510.
7. Item, C.B., Mihalek, I., Lichtarge, O., Jalan, A., Vodopiutz, J., Muhl, A. and Bodamer, O.A. 2007. Manifestation of hawkinsinuria in a patient compound heterozygous for hawkinsinuria and tyrosinemia III. Mol. Genet. Metab. 91: 379-383.
8. Online Mendelian Inheritance in Man, OMIM™. 2008. Johns Hopkins University, Baltimore, MD. MIM Number: 609695. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/

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: HPD (human) mapping to 12q24.31.

PRODUCT

HPPD (h): 293 Lysate represents a lysate of human HPPD transfected 293 cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

HPPD (h): 293 Lysate is suitable as a Western Blotting positive control for human reactive HPPD antibodies. Recommended use: 10-20 µl per lane.

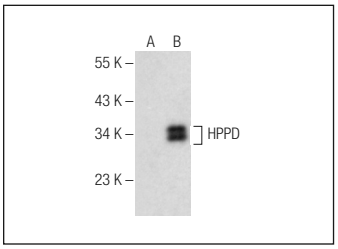
Control 293 Lysate: sc-110760 is available as a Western Blotting negative control lysate derived from non-transfected 293 cells.

HPPD (B-11): sc-390279 is recommended as a positive control antibody for Western Blot analysis of enhanced human HPPD expression in HPPD transfected 293 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



HPPD (B-11): sc-390279. Western blot analysis of HPPD expression in non-transfected: sc-110760 (**A**) and human HPPD transfected: sc-159724 (**B**) 293 whole cell lysates.

RESEARCH USE

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