SANTA CRUZ BIOTECHNOLOGY, INC.

E3BP (E-12): sc-377159

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

The pyruvate dehydrogenase (PDH) complex is a nuclear-encoded mitochondrial matrix enzyme complex that functions as the primary link between glycolysis and the tricarboxylic acid (TCA) cycle by catalyzing the irreversible conversion of pyruvate into acetyl-CoA. E3BP (E3-binding protein), also known as PDHX (pyruvate dehydrogenase protein X component) and lipoyl-containing pyruvate dehydrogenase complex component X, is a 501 amino acid mitochondrial protein that is required for anchoring E3 to the E2 core of the PDH complex, an event that is essential for a functional PDH complex. Defects in the gene encoding E3BP result in pyruvate dehydrogenase E3-binding protein deficiency, which is similar to PDH deficiency and Leigh syndrome in clinical presentation. Symptoms of E3BP deficiency can include lactic acidosis, delayed development, seizures, diplegia, cerebellar ataxia, optic atrophy, facial dysmorphism and episodic weakness.

REFERENCES

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2. Morava, E., et al. 2005. Mitochondrial dysfunction in a patient with Joubert syndrome. Neuropediatrics 36: 214-217.
3. Schiff, M., et al. 2006. Leigh's disease due to a new mutation in the PDHX gene. Ann. Neurol. 59: 709-714.
4. Brown, R.M., et al. 2006. Pyruvate dehydrogenase E3 binding protein (protein X) deficiency. Dev. Med. Child Neurol. 48: 756-760.
5. Smolle, M., et al. 2006. A new level of architectural complexity in the human pyruvate dehydrogenase complex. J. Biol. Chem. 281: 19772-19780.
6. McHugh, A., et al. 2006. PDC-E3BP is not a dominant T-cell autoantigen in primary biliary cirrhosis. Liver Int. 26: 406-413.
7. Miné, M., et al. 2006. A novel gross deletion caused by non-homologous recombination of the PDHX gene in a patient with pyruvate dehydrogenase deficiency. Mol. Genet. Metab. 89: 106-110.
8. Online Mendelian Inheritance in Man, OMIM™. 2008. Johns Hopkins University, Baltimore, MD. MIM Number: 608769. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
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CHROMOSOMAL LOCATION

Genetic locus: PDHX (human) mapping to 11p13.

SOURCE

E3BP (E-12) is a mouse monoclonal antibody raised against amino acids 151-280 mapping within an internal region of E3BP of human origin.

RESEARCH USE

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

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.

APPLICATIONS

E3BP (E-12) is recommended for detection of E3BP of 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).

Suitable for use as control antibody for E3BP siRNA (h): sc-77212, E3BP shRNA Plasmid (h): sc-77212-SH and E3BP shRNA (h) Lentiviral Particles: sc-77212-V.

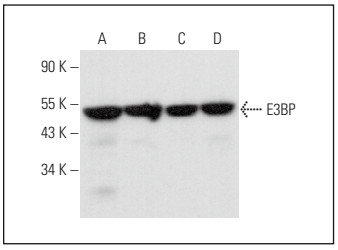
Molecular Weight of E3BP: 54 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, IMR-32 cell lysate: sc-2409 or JAR cell lysate: sc-2276.

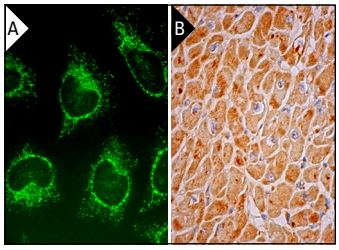
RECOMMENDED SUPPORT REAGENTS

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



E3BP (E-12): sc-377159. Western blot analysis of E3BP expression in Hep G2 (**A**), COLO 205 (**B**), IMR-32 (**C**) and JAR (**D**) whole cell lysates.



E3BP (E-12): sc-377159. Immunofluorescence staining of methanol-fixed HeLa cells showing mitochondrial localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human heart muscle tissue showing cytoplasmic staining of myocytes (**B**).

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.