SANTA CRUZ BIOTECHNOLOGY, INC.

E3BP (K-20): sc-79236

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

1. Robinson, B.H., et al. 1990. Defects in the E2 lipoyl transacetylase and the X-lipoyl containing component of the pyruvate dehydrogenase complex in patients with lactic acidemia. J. Clin. Invest. 85: 1821-1824.
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.

CHROMOSOMAL LOCATION

Genetic locus: PDHX (human) mapping to 11p13; Pdhx (mouse) mapping to 2 E2.

SOURCE

E3BP (K-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of E3BP 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-79236 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.

PROTOCOLS

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

APPLICATIONS

E3BP (K-20) is recommended for detection of E3BP 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).

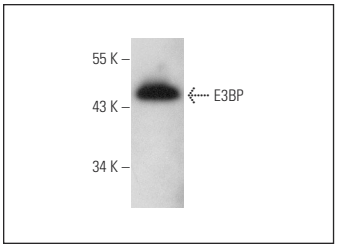
E3BP (K-20) is also recommended for detection of E3BP in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for E3BP siRNA (h): sc-77212, E3BP siRNA (m): sc-77213, E3BP shRNA Plasmid (h): sc-77212-SH, E3BP shRNA Plasmid (m): sc-77213-SH, E3BP shRNA (h) Lentiviral Particles: sc-77212-V and E3BP shRNA (m) Lentiviral Particles: sc-77213-V.

Molecular Weight of E3BP: 54 kDa.

Positive Controls: RT-4 whole cell lysate: sc-364257 or mouse heart extract: sc-2254.

DATA



E3BP (K-20): sc-79236. Western blot analysis of E3BP expression in RT-4 whole cell lysate.

SELECT PRODUCT CITATIONS

1. Koziel, A., et al. 2012. The influence of high glucose on the aerobic metabolism of endothelial EA.hy926 cells. Pflugers Arch. 464: 657-669.

RESEARCH USE

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