

## E3BP (S-19): sc-79238

### 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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### CHROMOSOMAL LOCATION

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

### SOURCE

E3BP (S-19) 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-79238 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### APPLICATIONS

E3BP (S-19) 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), 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 (S-19) 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: Mouse heart extract: sc-2254.

### RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

### STORAGE

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

### RESEARCH USE

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

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.