

PRPS1/2 (Y-16): sc-68130

BACKGROUND

PRPS (phosphoribosyl pyrophosphate synthetase) proteins catalyze the synthesis of phosphoribosyl pyrophosphate (PRPP). Three human PRPS isoforms exist and are encoded by three different genes. PRPS1 and PRPS2 (also known as PRS1 and PRS2, respectively) are ubiquitously expressed, while PRPS3 (also known as PRPS1L1) is specific to the testis. PRPP is an important substrate synthesized from MgATP and ribose-5-phosphate in a reaction that requires inorganic phosphate and magnesium as a cofactor. PRPP is essential in the synthesis of nearly all nucleotides, implying that PRPS1/2 play an important role in nucleotide biosynthesis and purine metabolism. A mutation in the gene encoding PRPS1 may result in PRPS superactivity, a disease characterized by gout and the overproduction of purine nucleotides, uric acid and PRPP. PRPS1 mutations can also lead to a reduction in PRPS1 activity resulting in ARTS syndrome or CMTX5 (Charcot-Marie-Tooth disease X-linked recessive type 5).

REFERENCES

1. Kunjara, S., et al. 1992. Phosphoribosyl pyrophosphate formation in the rat adrenal gland in relation to adrenal growth in experimental diabetes. *Diabetes* 41: 1429-1435.
2. Ishizuka, T., et al. 1992. Promoter regions of the human X-linked house-keeping genes PRPS1 and PRPS2 encoding phosphoribosylpyrophosphate synthetase subunit I and II isoforms. *Biochim. Biophys. Acta* 1130: 139-148.
3. Fujimori, S. 1996. PRPP synthetase superactivity. *Nippon Rinsho* 54: 3309-3314.
4. Ahmed, M., et al. 1999. Accelerated transcription of PRPS1 in X-linked overactivity of normal human phosphoribosylpyrophosphate synthetase. *J. Biol. Chem.* 274: 7482-7488.
5. García-Pavía, P., et al. 2003. Phosphoribosylpyrophosphate synthetase overactivity as a cause of uric acid overproduction in a young woman. *Arthritis Rheum.* 48: 2036-2041.
6. Tang, W., et al. 2006. Expression, purification, crystallization and preliminary X-ray diffraction analysis of human phosphoribosyl pyrophosphate synthetase 1 (PRS1). *Acta Crystallogr. Sect. F Struct. Biol. Cryst. Commun.* 62: 432-434.
7. de Brouwer, A.P., et al. 2007. Arts syndrome is caused by loss-of-function mutations in PRPS1. *Am. J. Hum. Genet.* 81: 507-518.
8. Kim, H.J., et al. 2007. Mutations in PRPS, which encodes the phosphoribosyl pyrophosphate synthetase enzyme critical for nucleotide biosynthesis, cause hereditary peripheral neuropathy with hearing loss and optic neuropathy (cmtx5). *Am. J. Hum. Genet.* 81: 552-558.
9. Li, S., et al. 2007. Crystal structure of human phosphoribosylpyrophosphate synthetase 1 reveals a novel allosteric site. *Biochem. J.* 401: 39-47.

SOURCE

PRPS1/2 (Y-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of PRPS1 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-68130 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

PRPS1/2 (Y-16) is recommended for detection of PRPS1, PRPS2 and PRPS1L1 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); also recommended for detection of PRPS3 of human origin and Prps13 of mouse origin.

PRPS1/2 (Y-16) is also recommended for detection of PRPS1, PRPS2 and PRPS1L1 in additional species, including equine, canine, bovine, porcine and avian.

Molecular Weight of PRPS1: 35 kDa

Molecular Weight of PRPS2: 34 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200.

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 or our catalog for detailed protocols and support products.