

PUS1 (B-6): sc-376443

BACKGROUND

PUS1 (pseudouridine synthase 1) belongs to the tRNA pseudouridine synthase truA family. PUS1 functions in the conversion of uridine into pseudouridine after the nucleotide has been incorporated into RNA. It may have a functional role in tRNAs and is also thought to assist in the peptidyl transfer reaction of rRNAs. As a nucleus-resident protein, PUS1 forms a complex with RARG and the SRA1 RNA. PUS1 is widely expressed, with highest levels of expression in the brain and skeletal muscle tissues. Defects in PUS1 are a cause of myopathy with lactic acidosis and sideroblastic anemia (MLASA), also known as mitochondrial myopathy and sideroblastic anemia. MLASA is a rare autosomal recessive oxidative phosphorylation disorder specific to bone marrow and skeletal muscle. The deduced human PUS1 protein contains 348 amino acids and shares 92% sequence homology with mouse PUS1.

REFERENCES

1. Arluison, V., et al. 1998. Transfer RNA-pseudouridine synthetase Pus1 of *Saccharomyces cerevisiae* contains one atom of zinc essential for its native conformation and tRNA recognition. *Biochemistry* 37: 7268-7276.
2. Arluison, V., et al. 1999. RNA:pseudouridine synthetase Pus1 from *Saccharomyces cerevisiae*: oligomerization property and stoichiometry of the complex with yeast tRNA(Phe). *Biochimie* 81: 751-756.
3. Arluison, V., et al. 1999. Pseudouridine synthetase Pus1 of *Saccharomyces cerevisiae*: kinetic characterisation, tRNA structural requirement and real-time analysis of its complex with tRNA. *J. Mol. Biol.* 289: 491-502.
4. Chen, J. and Patton, J.R. 1999. Cloning and characterization of a mammalian pseudouridine synthase. *RNA* 5: 409-419.
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6. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 608109. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
7. Patton, J.R., et al. 2003. *Caenorhabditis elegans* pseudouridine synthase 1 activity *in vivo*: tRNA is a substrate, but not U2 small nuclear RNA. *Biochem. J.* 372: 595-602.
8. Bykhovskaya, Y., et al. 2004. Missense mutation in pseudouridine synthase 1 (PUS1) causes mitochondrial myopathy and sideroblastic anemia (MLASA). *Am. J. Hum. Genet.* 74: 1303-1308.

CHROMOSOMAL LOCATION

Genetic locus: PUS1 (human) mapping to 12q24.33.

SOURCE

PUS1 (B-6) is a mouse monoclonal antibody raised against amino acids 148-427 mapping at the C-terminus of PUS1 of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

PUS1 (B-6) is recommended for detection of PUS1 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 PUS1 siRNA (h): sc-61417, PUS1 shRNA Plasmid (h): sc-61417-SH and PUS1 shRNA (h) Lentiviral Particles: sc-61417-V.

Molecular Weight of PUS1 isoform 1/2: 47/44 kDa.

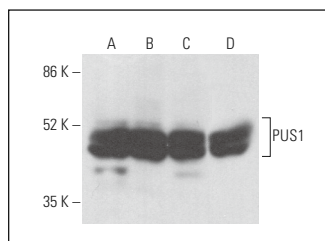
Positive Controls: HeLa whole cell lysate: sc-2200, A-431 whole cell lysate: sc-2201 or Jurkat whole cell lysate: sc-2204.

RECOMMENDED SUPPORT REAGENTS

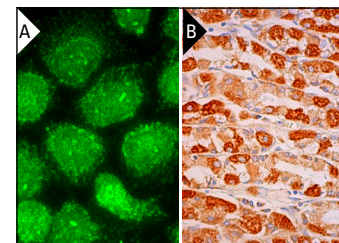
To ensure optimal results, the following support reagents are recommended:

- 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



PUS1 (B-6): sc-376443. Western blot analysis of PUS1 expression in HeLa (A), A-431 (B), Jurkat (C) and HEK293 (D) whole cell lysates.



PUS1 (B-6): sc-376443. Immunofluorescence staining of methanol-fixed HeLa cells showing nuclear localization (A). Immunoperoxidase staining of formalin fixed, paraffin-embedded human upper stomach tissue showing cytoplasmic staining of glandular cells (B).

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.