

Tim8A (N-20): sc-17052

BACKGROUND

The majority of mitochondrial-directed proteins are encoded by the nuclear genome and are transported to the mitochondria via regulated processes involving the mitochondrial Tom and Tim proteins. The mitochondrial Tim protein family is comprised of a large group of evolutionarily conserved proteins that are found in most eukaryotes. Import of nuclear-encoded precursor proteins into and across the mitochondrial inner membrane is mediated by two distinct complexes, the Tim23 complex and the Tim22 complex, which differ in their substrate specificity. Defects in Tim proteins are implicated in several neuro-degenerative diseases, suggesting important roles for Tim proteins in development and health. Tim8A and Tim8B, which map to human chromosomes Xq22.1 and 11q23.1-q23.2, respectively, are conserved proteins of the mitochondrial intermembrane space, which are organized in hetero-oligomeric complex with Tim13. Tim8A is highly expressed in fetal and adult brain. Tim8A is mutated in deafness dystonia syndrome, a novel type of disease that causes severe neurological defects, thought to be caused by a defective mitochondrial protein transport system.

REFERENCES

- Jin, H., et al. 1999. The human family of deafness/dystonia peptide (DDP) related mitochondrial import proteins. *Genomics* 61: 259-267.
- Bauer, M.F., et al. 1999. The mitochondrial Tim22 preprotein translocase is highly conserved throughout the eukaryotic kingdom. *FEBS Lett.* 464: 41-47.
- Rassow, J., et al. 1999. The preprotein translocase of the mitochondrial inner membrane: function and evolution. *J. Mol. Biol.* 286: 105-120.
- Koehler, C.M., et al. 1999. Human deafness dystonia syndrome is a mitochondrial disease. *Proc. Natl. Acad. Sci. USA* 96: 2141-2146.
- Paschen, S.A., et al. 2000. The role of the Tim8-13 complex in the import of Tim23 into mitochondria. *EMBO J.* 19: 6392-6400.
- Bauer, M.F. and Neupert, W. 2001. Import of proteins into mitochondria: a novel pathomechanism for progressive neurodegeneration. *J. Inher. Metab. Dis.* 24: 166-180.

CHROMOSOMAL LOCATION

Genetic locus: TIMM8A (human) mapping to Xq22.1; Timm8a1 (mouse) mapping to X E3.

SOURCE

Tim8A (N-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of Tim8A 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-17052 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

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

APPLICATIONS

Tim8A (N-20) is recommended for detection of Tim8A 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).

Tim8A (N-20) is also recommended for detection of Tim8A in additional species, including canine, bovine and porcine.

Suitable for use as control antibody for Tim8A siRNA (h): sc-41247, Tim8A siRNA (m): sc-41248, Tim8A shRNA Plasmid (h): sc-41247-SH, Tim8A shRNA Plasmid (m): sc-41248-SH, Tim8A shRNA (h) Lentiviral Particles: sc-41247-V and Tim8A shRNA (m) Lentiviral Particles: sc-41248-V.

Molecular Weight of Tim8A: 11 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.

SELECT PRODUCT CITATIONS

- Ravera, S., et al. 2009. Evidence for aerobic ATP synthesis in isolated myelin vesicles. *Int. J. Biochem. Cell Biol.* 41: 1581-1591.
- Ravera, S., et al. 2011. Characterization of myelin sheath F₀F₁-ATP synthase and its regulation by IF₁. *Cell Biochem. Biophys.* 59: 63-70.
- Ravera S., et al. 2011. Evidence for ectopic aerobic ATP production on C6 glioma cell plasma membrane. *Cell. Mol. Neurobiol.* 31: 313-321.
- Ravera, S., et al. 2013. Oxidative phosphorylation in sciatic nerve myelin and its impairment in a model of dysmyelinating peripheral neuropathy. *J. Neurochem.* 126: 82-92.

STORAGE

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



Try **Tim8A (T-40): sc-101282**, our highly recommended monoclonal alternative to Tim8A (N-20).