SANTA CRUZ BIOTECHNOLOGY, INC.

MPV17 (FL-176): sc-135219

BACKGROUND

MPV17, also known as SYM1, is a 176 amino acid mitochondrial inner membrane protein that belongs to the peroxisomal membrane protein PXMP2/4 family. MPV17 is expressed in pancreas, kidney, muscle, liver, lung, placenta, brain and heart. MPV17 plays an important role in regulating oxidative phosphorylation and mitochondrial DNA (mtDNA) maintenance. Mutations of MVP17 have been associated with the hepatocerebral form of mitochondrial DNA depletion syndrome (MDDS). MDDS is an autosomal recessive trait characterized by a reduction in mitochondrial DNA (mtDNA) copy number. MDDS may affect single organs, typically muscle or liver. Individuals with the hepatocerebral form of MDDS have early progressive liver failure and neurologic abnormalities, hypoglycemia and increased lactate in body fluids.

REFERENCES

1. Zwacka, R.M., et al. 1994. The glomerulosclerosis gene MPV17 encodes a peroxisomal protein producing reactive oxygen species. EMBO J. 13: 5129-5134.
2. Meyer zum Gottesberge, A.M., et al. 1996. Inner ear defect similar to Alport's syndrome in the glomerulosclerosis mouse model MPV17. Eur. Arch. Otorhinolaryngol. 253: 470-474.
3. Karadimas, C.L., et al. 2006. Navajo neurohepatopathy is caused by a mutation in the MPV17 gene. Am. J. Hum. Genet. 79: 544-548.
4. Wong, L.J., et al. 2007. Mutations in the MPV17 gene are responsible for rapidly progressive liver failure in infancy. Hepatology 46: 1218-1227.
5. Spinazzola, A., et al. 2008. Hepatocerebral form of mitochondrial DNA depletion syndrome: novel MPV17 mutations. Arch. Neurol. 65: 1108-1113.
6. Navarro-Sastre, A., et al. 2008. Lethal hepatopathy and leukodystrophy caused by a novel mutation in MPV17 gene: description of an alternative MPV17 spliced form. Mol. Genet. Metab. 94: 234-239.
7. Spinazzola, A., et al. 2008. Lack of founder effect for an identical mtDNA depletion syndrome (MDS)-associated MPV17 mutation shared by Navajos and Italians. Neuromuscul. Disord. 18: 315-318.
8. Viscomi, C., et al. 2009. Early-onset liver mtDNA depletion and late-onset proteinuric nephropathy in MPV17 knockout mice. Hum. Mol. Genet. 18: 12-26.

CHROMOSOMAL LOCATION

Genetic locus: MPV17 (human) mapping to 2p23.3; Mpv17 (mouse) mapping to 5 B1.

SOURCE

MPV17 (FL-176) is a rabbit polyclonal antibody raised against amino acids 1-176 representing full length MPV17 of mouse origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

MPV17 (FL-176) is recommended for detection of MPV17 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).

MPV17 (FL-176) is also recommended for detection of MPV17 in additional species, including equine, canine and bovine.

Suitable for use as control antibody for MPV17 siRNA (h): sc-94662, MPV17 siRNA (m): sc-149543, MPV17 shRNA Plasmid (h): sc-94662-SH, MPV17 shRNA Plasmid (m): sc-149543-SH, MPV17 shRNA (h) Lentiviral Particles: sc-94662-V and MPV17 shRNA (m) Lentiviral Particles: sc-149543-V.

Molecular Weight of MPV17: 20 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (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.