SANTA CRUZ BIOTECHNOLOGY, INC.

Wolframin (C-18): sc-47936

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

The Wolframin gene encodes a protein found in endoplasmic reticulum membrane of several tissues including brain, pancreas, lung and placenta. Loss-of-function mutations in both alleles result in Wolfram syndrome (also known as DIDMOAD, an autosomal recessive disorder that causes juvenile diabetes mellitus, diabetes insipidus, optic atrophy and a number of neurological symptoms including deafness, ataxia and peripheral neuropathy. A large number and variety of mutations in this gene, particularly in exon 8, can be associated with Wolfram syndrome. Mutations in this gene can also cause autosomal dominant deafness 6 (DFNA6), also known as DFNA14 or DFNA38.

REFERENCES

1. Osman, A.A., et al. 2003. Wolframin expression induces novel ion channel activity in endoplasmic reticulum membranes and increases intracellular calcium. J. Biol. Chem. 278: 52755-527562.
2. Larsen, Z.M., et al. 2004. Evidence for linkage on chromosome 4p16.1 in Type 1 diabetes Danish families and complete mutation scanning of the WFS1 (Wolframin) gene. Diabet. Med. 21: 218-222.
3. Smith, C.J., et al. 2004. Phenotype-genotype correlations in a series of Wolfram syndrome families. Diabetes Care 27: 2003-2009.
4. Yamaguchi, S., et al. 2004. Endoplasmic reticulum stress and N-glycosylation modulate protein. Biochem. Biophys. Res. Commun. 325: 250-256.
5. Fonseca, S.G., et al. 2005. WFS1 is a novel component of the unfolded protein response and maintains homeostasis of the endoplasmic reticulum in pancreatic β-cells. J. Biol. Chem. 280: 39609-39615.
6. Koido, K., et al. 2005. Polymorphisms in Wolframin (WFS1) gene are possibly related to for mood disorders. Int. J. Neuropsychopharmacol. 8: 235-244.
7. Philbrook, C., et al. 2005. Expressional and functional studies of Wolframin, the gene function deficient in Wolfram syndrome, in mice and patient cells. Exp. Gerontol. 40: 671-678.

CHROMOSOMAL LOCATION

Genetic locus: WFS1 (human) mapping to 4p16.1; Wfs1 (mouse) mapping to 5 B3.

SOURCE

Wolframin (C-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of Wolframin 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, ready P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

Wolframin (C-18) is recommended for detection of Wolframin 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), 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).

Wolframin (C-18) is also recommended for detection of Wolframin in additional species, including equine and canine.

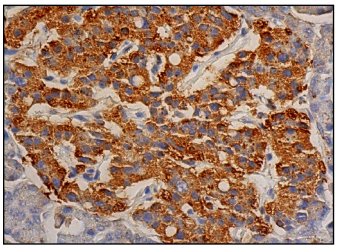
Suitable for use as control antibody for Wolframin siRNA (h): sc-61804, Wolframin siRNA (m): sc-61805, Wolframin shRNA Plasmid (h): sc-61804-SH, Wolframin shRNA Plasmid (m): sc-61805-SH, Wolframin shRNA (h) Lentiviral Particles: sc-61804-V and Wolframin shRNA (m) Lentiviral Particles: sc-61805-V.

Molecular Weight of Wolframin: 100 kDa.

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. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



Wolframin (C-18): sc-47936. Immunoperoxidase staining of formalin fixed, paraffin-embedded human pancreas tissue showing cytoplasmic staining of Islets of Langerhans.

SELECT PRODUCT CITATIONS

1. Goel, M., et al. 2011. Cochlin induced TREK-1 co-expression and annexin A2 secretion: role in trabecular meshwork cell elongation and motility. PLoS ONE 6: e23070.
2. Elli, F.M., et al. 2012. A new structural rearrangement associated to Wolfram syndrome in a child with a partial phenotype. Gene 509: 168-172.

RESEARCH USE

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