SANTA CRUZ BIOTECHNOLOGY, INC.

NUBPL (G-7): sc-393245

BACKGROUND

NUBPL (nucleotide-binding protein-like) is a 319 amino acid mitochondrial protein that belongs to the Mrp/NBP35 ATP-binding protein family and exists as two alternatively spliced isoforms. Required for the assembly of the mitochondrial membrane respiratory chain NADH dehydrogenase (complex I), NUBPL may deliver one or more Fe-S clusters to complex I subunits. With highest expression in liver and kidney, NUBPL is expressed at significant levels in small intestine and brain. Defects in NUBPL are a cause of mitochondrial complex I deficiency (MT-C1D), a disorder of the mitochondrial respiratory chain that causes a wide range of afflictions from lethal neonatal disease to adult-onset neurodegenerative disorders. Phenotypes include macrocephaly with progressive leukodystrophy, non-specific encephalopathy, cardiomyopathy, myopathy, liver disease, Leigh syndrome, Leber hereditary optic neuropathy and some forms of Parkinson disease.

REFERENCES

1. Online Mendelian Inheritance in Man, OMIM™. 1987. Johns Hopkins University, Baltimore, MD. MIM Number: 252010. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
2. Larsson, N.G., et al. 1991. Leber's hereditary optic neuropathy and complex I deficiency in muscle. Ann. Neurol. 30: 701-708.
3. Robinson, B.H. 1998. Human complex I deficiency: clinical spectrum and involvement of oxygen free radicals in the pathogenicity of the defect. Biochim. Biophys. Acta 1364: 271-286.
4. Smeitink, J. and van den Heuvel, L. 1999. Human mitochondrial complex I in health and disease. Am. J. Hum. Genet. 64: 1505-1510.
5. Triepels, R.H., et al. 2001. Respiratory chain complex I deficiency. Am. J. Med. Genet. 106: 37-45.
6. Tretter, L., et al. 2004. Initiation of neuronal damage by complex I deficiency and oxidative stress in Parkinson's disease. Neurochem. Res. 29: 569-577.
7. Sheftel, A.D., et al. 2009. Human ind1, an iron-sulfur cluster assembly factor for respiratory complex I. Mol. Cell. Biol. 29: 6059-6073.

CHROMOSOMAL LOCATION

Genetic locus: NUBPL (human) mapping to 14q12.

SOURCE

NUBPL (G-7) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 140-163 within an internal region of NUBPL of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-393245 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

NUBPL (G-7) is recommended for detection of NUBPL 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for NUBPL siRNA (h): sc-92266, NUBPL shRNA Plasmid (h): sc-92266-SH and NUBPL shRNA (h) Lentiviral Particles: sc-92266-V.

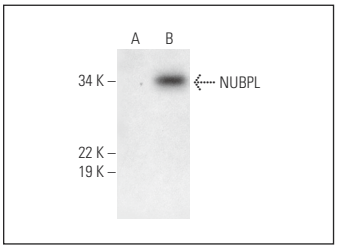
Molecular Weight of NUBPL isoforms: 34/18 kDa.

Positive Controls: NUBPL (h): 293T Lysate: sc-113739.

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.

DATA



NUBPL (G-7): sc-393245. Western blot analysis of NUBPL expression in non-transfected: sc-117752 (**A**) and human NUBPL transfected: sc-113739 (**B**) 293T whole cell lysates.

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 for detailed protocols and support products.