

# NUBPL (E-12): sc-137644

## 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 2 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

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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., et al. 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.
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## CHROMOSOMAL LOCATION

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

## SOURCE

NUBPL (E-12) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping within an internal region of NUBPL of human origin.

## PRODUCT

Each vial contains 100 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-137644 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

NUBPL (E-12) is recommended for detection of NUBPL of human origin by Western Blotting (starting dilution 1:100, dilution range 1:50-1:500), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:25, dilution range 1:25-1:250) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); non cross-reactive with NUBP1 or NUBP2.

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.

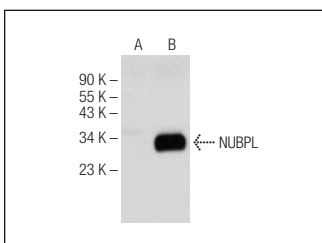
Molecular Weight of NUBPL isoforms: 34/18 kDa.

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

## RECOMMENDED 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.

## DATA



NUBPL (E-12): sc-137644. 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.

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Try **NUBPL (G-11): sc-398217** or **NUBPL (G-7): sc-393245**, our highly recommended monoclonal alternatives to NUBPL (E-12).