

# KIAA0391 (I-14): sc-139152

## BACKGROUND

KIAA0391, also known as MRPP3 (mitochondrial ribonuclease P protein 3), is a 583 amino acid protein that is located in the mitochondria. KIAA0391 functions in tRNA maturation and is a component of mitochondrial ribonuclease P which cleaves tRNA at the 5'-end. Mitochondrial ribonuclease P is an enzyme composed of RG9MTD1, MRPP22 and KIAA0391 subunits. Due to alternative splicing events, KIAA0391 exists as four isoforms. The gene encoding KIAA0391 maps to human chromosome 14q13.2, which contains about 700 genes and makes up about 3.5% of human cellular DNA. Chromosome 14 encodes the presenilin 1 (PSEN1) gene, which is one of the three key genes associated with the development of Alzheimer's disease. The SERPINA1 gene is located on chromosome 14 and when defective leads to the genetic disorder  $\alpha$ 1-antitrypsin deficiency. This disorder is characterized by severe lung complications and liver dysfunction. Notably, the immunoglobulin heavy chain locus is found on chromosome 14 and is suggested to fuse with the chromosome 19 encoded protein BCL3 by (14;19) translocation events in a variety of B cell malignancies.

## REFERENCES

1. Nagase, T., et al. 1997. Prediction of the coding sequences of unidentified human genes. VII. The complete sequences of 100 new cDNA clones from brain which can code for large proteins *in vitro*. DNA Res. 4: 141-150.
2. Heilig, R., et al. 2003. The DNA sequence and analysis of human chromosome 14. Nature 421: 601-607.
3. Godbolt, A.K., et al. 2004. A presenilin 1 R278I mutation presenting with language impairment. Neurology 63: 1702-1704.
4. Stolk, J., et al. 2006.  $\alpha$ 1-antitrypsin deficiency: current perspective on research, diagnosis, and management. Int. J. Chron. Obstruct. Pulmon. Dis. 1: 151-160.
5. Vetrivel, K.S., et al. 2006. Pathological and physiological functions of presenilins. Mol. Neurodegener. 1: 4.
6. Albani, D., et al. 2007. Presenilin-1 mutation E318G and familial Alzheimer's disease in the Italian population. Neurobiol. Aging 28: 1682-1688.
7. Cruz, P.E., et al. 2007. The promise of gene therapy for the treatment of  $\alpha$ -1 antitrypsin deficiency. Pharmacogenomics 8: 1191-1198.

## CHROMOSOMAL LOCATION

Genetic locus: KIAA0391 (human) mapping to 14q13.2; 1110008L16Rik (mouse) mapping to 12 C1.

## SOURCE

KIAA0391 (I-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of KIAA0391 of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-139152 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

KIAA0391 (I-14) is recommended for detection of KIAA0391 of human origin and 1110008L16Rik of mouse origin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g 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); non cross-reactive with other KIAA family members .

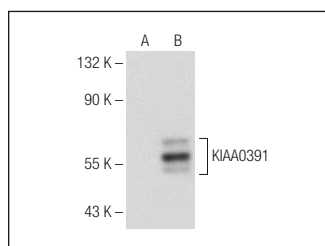
KIAA0391 (I-14) is also recommended for detection of KIAA0391 in additional species, including equine.

Suitable for use as control antibody for KIAA0391 siRNA (h): sc-92117, 1110008L16Rik siRNA (m): sc-108150, KIAA0391 shRNA Plasmid (h): sc-92117-SH, 1110008L16Rik shRNA Plasmid (m): sc-108150-SH, KIAA0391 shRNA (h) Lentiviral Particles: sc-92117-V and 1110008L16Rik shRNA (m) Lentiviral Particles: sc-108150-V.

Molecular Weight of KIAA0391: 25/56/65/67 kDa.

Positive Controls: KIAA0391 (h): 293T Lysate: sc-114148.

## DATA



KIAA0391 (I-14): sc-139152. Western blot analysis of KIAA0391 expression in non-transfected: sc-117752 (A) and human KIAA0391 transfected: sc-114148 (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](http://www.scbt.com) or our catalog for detailed protocols and support products.



Try **KIAA0391 (D-2): sc-390380** or **KIAA0391 (C-4): sc-514705**, our highly recommended monoclonal alternatives to KIAA0391 (I-14).