

# PANK2 (4C11): sc-100405

## BACKGROUND

PANK2 (pantothenate kinase 2), also known as HSS, HARP, PKAN or NBIA1, is a ubiquitously expressed 570 amino acid member of the pantothenate kinase family of enzymes that are involved in the synthesis of coenzyme A (CoA). Localized to the cytoplasm and mitochondria, PANK2 is thought to be the chief regulator of CoA biosynthesis, catalyzing the first of five steps in the biosynthetic pathway. Regulated by feedback inhibition from synthesized CoA, PANK2 catalyzes the ATP-dependent conversion of pantothenate to 4'-phosphopantothenate, thus initiating the first committed step in CoA biosynthesis. Defects in the gene encoding PANK2 are the cause of pantothenate kinase-associated neurodegeneration (PKAN) and hypoprebetalipoproteinemia, acanthocytosis, retinitis pigmentosa and pallidal degeneration (HARP). PKAN and HARP are rare disorders characterized by extrapyramidal dysfunction and progressive dementia, both of which are caused by an accumulation of iron in the brain. PANK2 is expressed as three isoforms due to alternative splicing events.

## REFERENCES

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2. Ching, K.H., et al. 2002. HARP syndrome is allelic with pantothenate kinase-associated neurodegeneration. *Neurology* 58: 1673-1674.
3. Hörtnagel, K., et al. 2003. An isoform of hPANK2, deficient in pantothenate kinase-associated neurodegeneration, localizes to mitochondria. *Hum. Mol. Genet.* 12: 321-327.
4. Hartig, M.B., et al. 2006. Genotypic and phenotypic spectrum of PANK2 mutations in patients with neurodegeneration with brain iron accumulation. *Ann. Neurol.* 59: 248-256.
5. Kazek, B., et al. 2007. A novel PANK2 gene mutation: clinical and molecular characteristics of patients short communication. *J. Child Neurol.* 22: 1256-1259.
6. Leonardi, R., et al. 2007. Localization and regulation of mouse pantothenate kinase 2. *FEBS Lett.* 581: 4639-4644.
7. Saleheen, D., et al. 2007. Novel mutation in the PANK2 gene leads to pantothenate kinase-associated neurodegeneration in a Pakistani family. *Pediatr. Neurol.* 37: 296-298.
8. Hong, B.S., et al. 2007. Crystal structures of human pantothenate kinases. Insights into allosteric regulation and mutations linked to a neurodegeneration disorder. *J. Biol. Chem.* 282: 27984-27993.
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## CHROMOSOMAL LOCATION

Genetic locus: PANK2 (human) mapping to 20p13.

## STORAGE

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

## SOURCE

PANK2 (4C11) is a mouse monoclonal antibody raised against recombinant PANK2 of human origin.

## PRODUCT

Each vial contains 50 µg IgG<sub>2a</sub> in 500 µl of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

PANK2 (4C11) is recommended for detection of PANK2 of human origin by immunofluorescence and 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).

Suitable for use as control antibody for PANK2 siRNA (h): sc-76042, PANK2 shRNA Plasmid (h): sc-76042-SH and PANK2 shRNA (h) Lentiviral Particles: sc-76042-V.

Molecular Weight of PANK2 isoform 1: 63 kDa.

Molecular Weight of PANK2 isoform 2: 59 kDa.

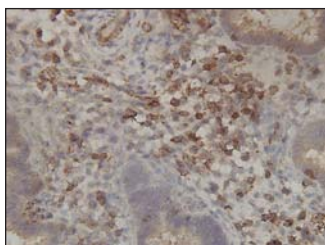
Molecular Weight of PANK2 isoform 3: 48 kDa.

Positive Controls: human endometrium tissue.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Immunofluorescence: use goat anti-mouse IgG-FITC: sc-2010 (dilution range: 1:100-1:400) or goat anti-mouse IgG-TR: sc-2781 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941. 2) Immunohistochemistry: use ImmunoCruz™: sc-2050 or ABC: sc-2017 mouse IgG Staining Systems.

## DATA



PANK2 (4C11): sc-100405. Immunoperoxidase staining of formalin-fixed, paraffin-embedded human endometrium tissue showing cytoplasmic localization.

## 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.