SunLong Biotech Co.,LTD
Tel: 0086-571- 56623320 Fax:0086-571- 56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

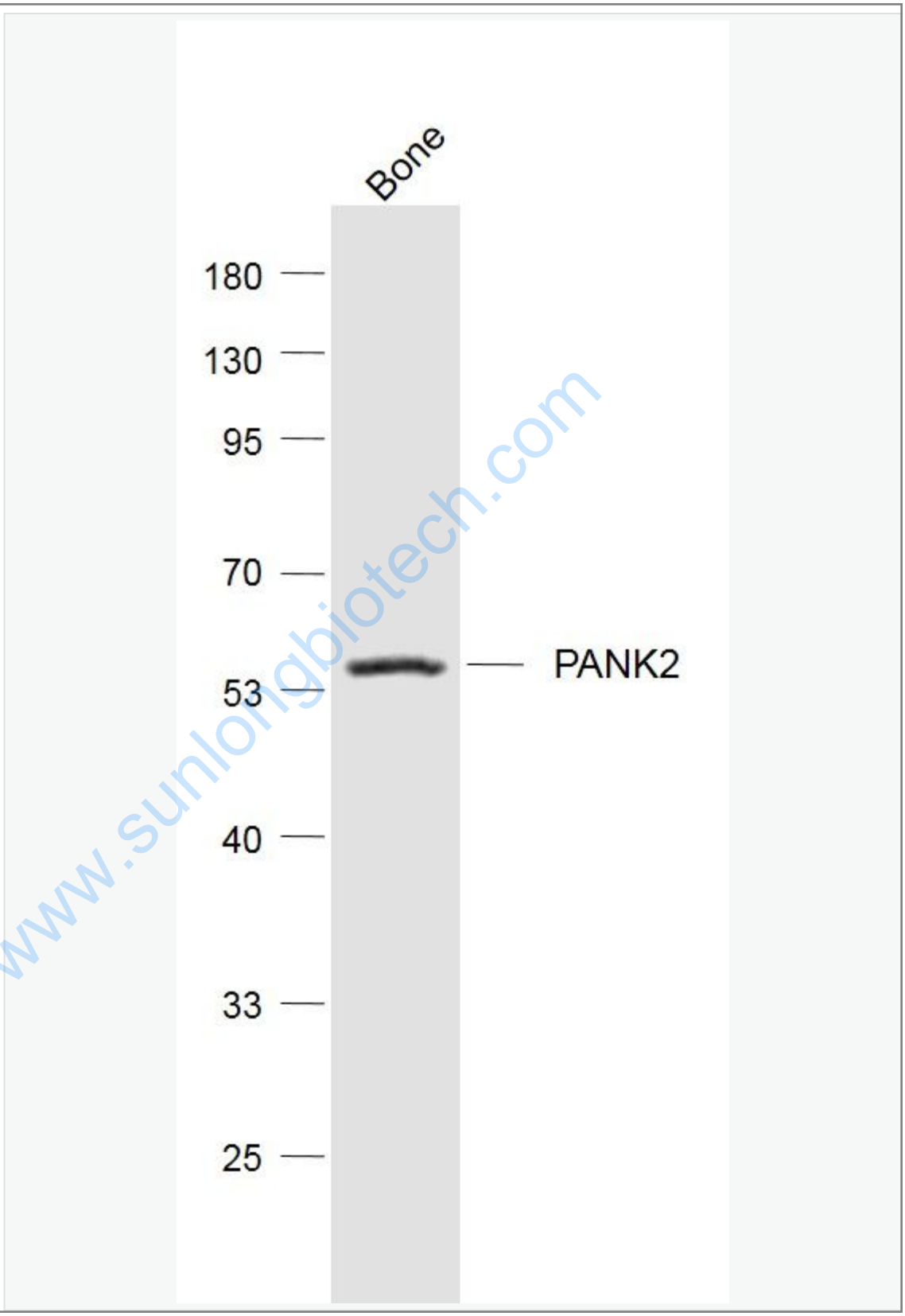
Rabbit Anti-PANK2 antibody

SL8338R

Product Name:	PANK2
Chinese Name:	泛酸激酶2抗体
Alias:	C20orf48; HARP; hPANK2; HSS; MGC15053; NBIA1; PANK2; PANK2_HUMAN; Pantothenate kinase 2 (Hallervorden Spatz syndrome); Pantothenate kinase 2; Pantothenic acid kinase 2; PKAN; RP23 387C21.4.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:50-200（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	57kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human PANK2:401-500/570
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Defects in PANK2 are the cause of neurodegeneration with brain iron accumulation type 1 (NBIA1); also known as pantothenate kinase-associated neurodegeneration (PKAN) or Hallervorden-Spatz syndrome (HSS). It is an autosomal recessive neurodegenerative disorder associated with iron accumulation in the brain, primarily in the basal ganglia. Clinical manifestations include progressive muscle spasticity,

	hyperreflexia, muscle rigidity, dystonia, dysarthria, and intellectual deterioration which progresses to severe dementia over several years. It is clinically classified into classic, atypical, and intermediate phenotypes. Classic forms present with onset in the first decade, rapid progression, loss of independent ambulation within 15 years. Atypical forms have onset in the second decade, slow progression, maintenance of independent ambulation up to 40 years later. Intermediate forms manifest onset in the first decade with slow progression or onset in the second decade with rapid progression. Patients with early onset tend to also develop pigmentary retinopathy, whereas those with later onset tend to also have speech disorders and psychiatric features. All patients have the 'eye of the tiger' sign on brain MRI. Defects in PANK2 are the cause of hypoprebetalipoproteinemia, acanthocytosis, retinitis pigmentosa, and pallidal degeneration (HARP). HARP is a rare syndrome with many clinical similarities to NBIA1. **Function:** May be the master regulator of the CoA biosynthesis (By similarity). **Subcellular Location:** Isoform 1: Mitochondrion. Isoform 2: Cytoplasm (Potential). **Tissue Specificity:** Ubiquitous. **DISEASE:** Defects in PANK2 are the cause of neurodegeneration with brain iron accumulation type 1 (NBIA1) [MIM:234200]; also known as pantothenate kinase-associated neurodegeneration (PKAN) or Hallervorden-Spatz syndrome (HSS). It is an autosomal recessive neurodegenerative disorder associated with iron accumulation in the brain, primarily in the basal ganglia. Clinical manifestations include progressive muscle spasticity, hyperreflexia, muscle rigidity, dystonia, dysarthria, and intellectual deterioration which progresses to severe dementia over several years. It is clinically classified into classic, atypical, and intermediate phenotypes. Classic forms present with onset in the first decade, rapid progression, loss of independent ambulation within 15 years. Atypical forms have onset in the second decade, slow progression, maintenance of independent ambulation up to 40 years later. Intermediate forms manifest onset in the first decade with slow progression or onset in the second decade with rapid progression. Patients with early onset tend to also develop pigmentary retinopathy, whereas those with later onset tend to also have speech disorders and psychiatric features. All patients have the 'eye of the tiger' sign on brain MRI. Defects in PANK2 are the cause of hypoprebetalipoproteinemia, acanthocytosis, retinitis pigmentosa, and pallidal degeneration (HARP) [MIM:607236]. HARP is a rare syndrome with many clinical similarities to NBIA1. **Similarity:** Belongs to the type II pantothenate kinase family.

	SWISS: Q9BZ23 **Gene ID:** 80025 **Database links:** Entrez Gene: 80025Human Omim: 606157Human SwissProt: Q9BZ23Human Unigene: 516859Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:	

	Sample: Bone (Mouse) Lysate at 40 ug Primary: Anti-PANK2 (SL8338R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 57 kD Observed band size: 57 kD