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

Rabbit Anti-SMUBP2 antibody

SL11757R

Product Name:	SMUBP2
Chinese Name:	免疫球蛋白μ链Binding protein2抗体
Alias:	AEP; Antifreeze enhancer binding protein; ATP-dependent helicase IGHMBP2; Cardiac transcription factor 1; Cardiac transcription factor1; CATF 1; CATF1; DNA-binding protein SMUBP-2; GF-1; Glial factor 1; HCSA; HMN 6; HMN6; IGHMBP 2; Ighmbp2; Immunoglobulin mu binding protein 2; Immunoglobulin mu binding protein2; Immunoglobulin mu-binding protein 2; Immunoglobulin S mu binding protein 2; Immunoglobulin S mu binding protein2; RIPE3 b1; RIPE3b 1; RIPE3b1; SMARD 1; SMARD1; SMBP2_HUMAN; SMUBP 2.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Pig,Cow,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	109kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human SMUBP2:271-355/993
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:	IGHMBP2 is a 993 amino acid nuclear and cytoplasmic protein that is ubiquitously

	expressed. Belonging to the DNA2/NAM7 helicase family, IGHMBP2 is a 5' to 3' helicase that unwinds RNA and DNA duplexes in an ATP-dependent reaction. IGHMBP2 also acts as a transcriptional regulator and is necessary for transcriptional activation of the flounder liver-type antifreeze protein gene. IGHMBP2 exists as a homooligomer and is part of the cytosolic ribonucleoprotein complex. Mutations in the gene encoding IGHMBP2 are suggested to lead to distal hereditary motor neuronopathy type 6 (HMN6), also known as spinal muscular atrophy distal autosomal recessive 1 (DSMA1) or spinal muscular atrophy with respiratory distress 1 (SMARD1). HMN6 is characterized by weakness and wasting of distal muscles of the anterior tibial and peroneal compartments of the legs and severe respiratory distress. **Function:** 5' to 3' helicase that unwinds RNA and DNA duplices in an ATP-dependent reaction. Acts as a transcription regulator. Required for the transcriptional activation of the flounder liver-type antifreeze protein gene. Exhibits strong binding specificity to the enhancer element B of the flounder antifreeze protein gene intron. Binds to the insulin II gene RIPE3B enhancer region. May be involved in translation (By similarity). DNA-binding protein specific to 5'-phosphorylated single-stranded guanine-rich sequence related to the immunoglobulin mu chain switch region. Preferentially binds to the 5'-GGGCT-3' motif. Interacts with tRNA-Tyr. Stimulates the transcription of the human neurotropic virus JCV. **Subunit:** Homooligomer. Interacts with RUVBL1, RUVBL2, GTF3C1 andABT1. Is part of large cytosolic ribonucleoprotein complexes(Probable). Associates with the ribosomes. **Subcellular Location:** Nucleus. Cytoplasm. **Tissue Specificity:** Expressed in all tissues examined. **Post-translational modifications:** Phosphorylated upon DNA damage, probably by ATM or ATR. **DISEASE:** Defects in IGHMBP2 are the cause of distal hereditary motor neuronopathy type 6 (HMN6) [MIM:604320]; also known as spinal muscular atrophy distal autosomal recessive 1 (DSMA1) or spinal muscular atrophy with respiratory distress 1 (SMARD1). Distal hereditary motor neuronopathies constitute a heterogeneous group of neuromuscular disorders caused by selective degeneration of motor neurons in the anterior horn of the spinal cord, without sensory deficit in the posterior horn. The overall clinical picture consists of a classical distal muscular atrophy syndrome in the legs without clinical sensory loss. The disease starts with weakness and wasting of distal muscles of the anterior tibial and peroneal compartments of the legs. Later on, weakness and atrophy may expand to the proximal muscles of the lower limbs and/or to the distal

upper limbs. The most prominent symptoms of HMN6 are severe respiratory distress resulting from diaphragmatic paralysis with eventration shown on chest x-ray and predominant involvement of the upper limbs and distal muscles.

Similarity:
Belongs to the DNA2/NAM7 helicase family.
Contains 1 AN1-type zinc finger.
Contains 1 R3H domain.

SWISS:
P38935

Gene ID:
3508

Database links:

Entrez Gene: 3508Human

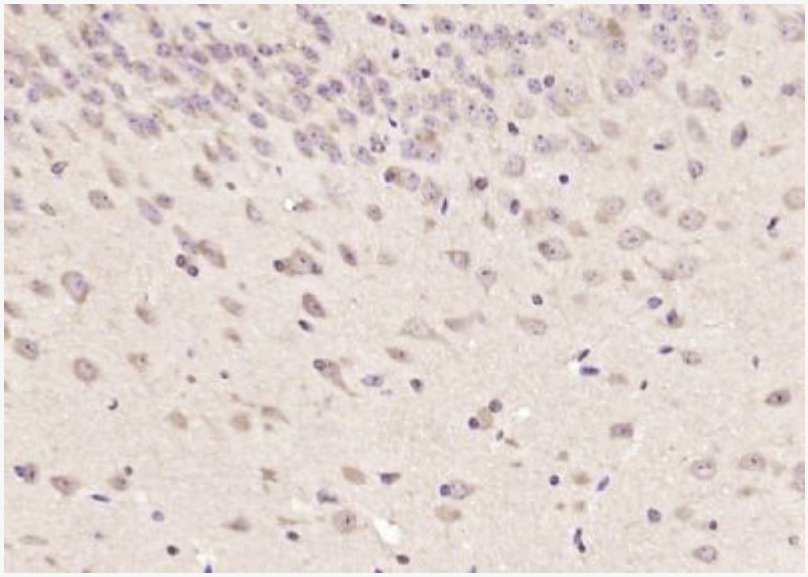
Omim: 600502Human

SwissProt: P38935Human

Unigene: 503048Human

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:



	Paraformaldehyde-fixed, paraffin embedded (mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (SMUBP2) Polyclonal Antibody, Unconjugated (SL11757R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.