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

Rabbit Anti-HSPC014/C13orf12 antibody

SL18086R

Product Name:	HSPC014/C13orf12
Chinese Name:	13号染色体开放阅读框12抗体
Alias:	2510048O06Rik; C13orf12; Chromosome 13 open reading frame 12; HSPC 014; HSPC036 protein; hUMP 1; hUMP1; PNAS 110; PNAS110; Pomp; POMP_HUMAN; Proteasome maturation protein; Proteassemblin; Protein UMP1 homolog; UMP 1; UMP1; UMP1, yeast, homolog of; Voltage gated K channel beta subunit 4.1; Voltage-gated K channel beta subunit 4.1; voltage-gated potassium channel beta subunit 4.1.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,
Applications:	ELISA=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:	16kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human HSPC014/C13orf12:3-100/141
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:	The protein encoded by this gene is a molecular chaperone that binds 20S preproteasome components and is essential for 20S proteasome formation. The 20S proteasome is the proteolytically active component of the 26S proteasome complex.

The encoded protein is degraded before the maturation of the 20S proteasome is complete. A variant in the 5' UTR of this gene has been associated with KLICK syndrome, a rare skin disorder.[provided by RefSeq, Aug 2010]

Function:
Molecular chaperone essential for the assembly of standard proteasomes and immunoproteasomes. Degraded after completion of proteasome maturation. Mediates the association of 20S preproteasome with the endoplasmic reticulum.

Subcellular Location:
Cytoplasm > cytosol. Nucleus. Microsome membrane.

Tissue Specificity:
Strongly expressed from the basal layer to the granular layer of healthy epidermis, whereas in KLICK patients there is a gradual decrease of expression toward the granular layer.

DISEASE:
Defects in POMP are the cause of keratosis linearis with ichthyosis congenita and sclerosing keratoderma (KLICK) [MIM:601952]. KLICK is a keratinizing disorder characterized by ichthyosis, palmoplantar keratoderma with constricting bands around fingers, flexural deformities of fingers and keratotic papules in a linear distribution on the flexural side of large joints. Histological examination of the skin of affected individuals shows hypertrophy and hyperplasia of the spinous, granular and horny epidermal layer.

Similarity:
Belongs to the POMP/UMP1 family.

SWISS:
Q9Y244

Gene ID:
51371

Database links:

Entrez Gene: 51371 Human

Entrez Gene: 66537 Mouse

Entrez Gene: 288455 Rat

Omim: 613386 Human

SwissProt: Q9Y244 Human

	SwissProt: Q9CQT5 Mouse Unigene: 268742 Human Unigene: 332855 Mouse Unigene: 28242 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.