13 号染色体开放阅读框 12 抗体

产品货号： mlR18086

英文名称： HSPC014/C13orf12

中文名称： 13 号染色体开放阅读框 12 抗体

别　　名： 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.

研究领域： 细胞生物　免疫学　信号转导

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 16kDa

细胞定位： 细胞核 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human HSPC014/C13orf12:3-100/141

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： 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

Important Note:

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