KBTBD13 蛋白抗体

产品货号： mlR17086

英文名称： KBTBD13

中文名称： KBTBD13 蛋白抗体

别　　名： 5430433E21Rik; HCG1645727; KBTBD_HUMAN; Kbtbd13; Kelch repeat and BTB (POZ) domain containing 13; Kelch repeat and BTB domain-containing protein 13; NEM6.

研究领域： 细胞生物　免疫学　发育生物学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Cow,

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： 49kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human KBTBD13:381-458/458

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 gene belongs to a family of genes encoding proteins containing a BTB domain and several kelch repeats. The BTB domain functions as a protein-protein interaction module, which includes an ability to self-associate or to interact with non-BTB domain-containing proteins. The kelch motif typically occurs in groups of five to seven repeats, and has been found in proteins with diverse functions. Known functions of these family members include transcription regulation, ion channel tetramerization and gating, protein ubiquitination or degradation, and cytoskeleton regulation. The exact function of this family member has yet to be determined. [provided by RefSeq, Jun 2010]

Subcellular Location:

Cytoplasm.

Tissue Specificity:

Expressed in skeletal muscle.

DISEASE:

Defects in KBTBD13 are the cause of nemaline myopathy type 6 (NEM6) [MIM:609273]. A form of nemaline myopathy characterized by childhood onset of slowly progressive proximal muscle weakness, exercise intolerance, and slow movements with stiff muscles. Patients are unable to run or correct themselves from falling over.

Similarity:

Contains 1 BTB (POZ) domain.

Contains 5 Kelch repeats.

SWISS:

C9JR72

Important Note:

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