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KBTBD13 Rabbit pAb

Catalog # AP97965

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	C9JR72
Other Accession	C9JR72
Predicted	Human, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	49485
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human KBTBD13
Epitope Specificity	381-458/458
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 BTB (POZ) domain. Contains 5 Kelch repeats.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Gene ID	390594
Other Names	HCG1645727; NEM6; KBTBD_HUMAN; KBTBD13; kelch repeat and BTB domain containing 13; kelch repeat and BTB (POZ) domain containing 13; nemaline myopathy type 6
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000

Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	KBTBD13
Function	Substrate-specific adapter of a BCR (BTB-CUL3-RBX1) E3 ubiquitin ligase complex.
Cellular Location	Cytoplasm
Tissue Location	Expressed in skeletal muscle.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.