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

Rabbit Anti-MVK antibody

SL20155R

Product Name:	MVK
Chinese Name:	甲羟戊酸激酶抗体
Alias:	LH receptor mRNA binding protein; LRBP; Mevalonate kinase; Mevalonic aciduria; MK antibody MVLK; KIME_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,
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:	42kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human MVK:101-200/396
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	Preservative: 15mM Sodium Azide, Constituents: 1% BSA, 0.01M PBS, pH 7.4
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:	MVK encodes the peroxisomal enzyme mevalonate kinase. Mevalonate is a key intermediate, and mevalonate kinase a key early enzyme, in isoprenoid and sterol synthesis. Mevalonate kinase deficiency caused by mutation of MVK results in mevalonic aciduria. **Function:**

May be a regulatory site in cholesterol biosynthetic pathway.

Subunit:
Homodimer.

Subcellular Location:
Cytoplasm.

DISEASE:
Defects in MVK are the cause of mevalonic aciduria (MEVA) [MIM:610377]. It is an accumulation of mevalonic acid which causes a variety of symptoms such as psychomotor retardation, dysmorphic features, cataracts, hepatosplenomegaly, lymphadenopathy, anemia, hypotonia, myopathy, and ataxia.
Defects in MVK are the cause of hyperimmunoglobulinemia D and periodic fever syndrome (HIDS) [MIM:260920]. HIDS is an autosomal recessive disease characterized by recurrent episodes of unexplained high fever associated with skin rash, diarrhea, adenopathy (swollen, tender lymph nodes), athralgias and/or arthritis. Concentration of IgD, and often IgA, are above normal.

Similarity:
Belongs to the GHMP kinase family. Mevalonate kinase subfamily.

SWISS:
Q03426

Gene ID:
4598

Database links:

Entrez Gene: 4598Human

Omim: 251170Human

SwissProt: Q03426Human

Unigene: 130607Human

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