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Rabbit Anti-MCFD2 antibody

SL18724R

Product Name:	MCFD2
Chinese Name:	多种凝血因子缺乏蛋白2抗体
Alias:	1810021C21Rik; DKFZp686G21263; F5F8D; LMAN1IP; MCFD 2; Mcfd2; MCFD2_HUMAN; Multiple coagulation factor deficiency protein 2; Neural stem cell derived neuronal survival protein; Neural stem cell-derived neuronal survival protein; SDNSF.
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:	16kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human MCFD2:1-100/146
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:	This gene encodes a soluble luminal protein with two calmodulin-like EF-hand motifs at its C-terminus. This protein forms a complex with LAMN1 (lectin mannose binding protein 1; also known as ERGIC-53) that facilitates the transport of coagulation factors V (FV) and VIII (FVIII) from the endoplasmic reticulum to the Golgi apparatus via an

endoplasmic reticulum Golgi intermediate compartment (ERGIC). Mutations in this gene cause combined deficiency of FV and FVIII (F5F8D); a rare autosomal recessive bleeding disorder characterized by mild to moderate bleeding and coordinate reduction in plasma FV and FVIII levels. This protein has also been shown to maintain stem cell potential in adult central nervous system and is a marker for testicular germ cell tumors. The 3' UTR of this gene contains a transposon-like human repeat element named 'THE 1'. A processed RNA pseudogene of this gene is on chromosome 6p22.1. Alternative splicing results in multiple transcript variants encoding distinct isoforms. [provided by RefSeq, Jun 2010]

Function:
The MCFD2-LMAN1 complex forms a specific cargo receptor for the ER-to-Golgi transport of selected proteins. Plays a role in the secretion of coagulation factors.

Subcellular Location:
Endoplasmic reticulum-Golgi intermediate compartment. Endoplasmic reticulum. Golgi apparatus.

DISEASE:
Defects in MCFD2 are a cause of factor V and factor VIII combined deficiency type 2 (F5F8D2) [MIM:613625]; also known as multiple coagulation factor deficiency 2 (MCFD2). F5F8D2 is a blood coagulation disorder characterized by bleeding symptoms similar to those in hemophilia or parahemophilia, that are caused by single deficiency of FV or FVIII, respectively. The most common symptoms are epistaxis, menorrhagia, and excessive bleeding during or after trauma. Plasma levels of coagulation factors V and VIII are in the range of 5 to 30% of normal.

Similarity:
Contains 2 EF-hand domains.

SWISS:
Q8NI22

Gene ID:
90411

Database links:

Entrez Gene: 90411 Human

Omim: 607788 Human

SwissProt: Q8NI22 Human

Unigene: 293689 Human

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