

MCFD2 Rabbit pAb

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Catalog # AP94135

Product Information

Application	WB, IHC-P, IHC-F, IF
Reactivity	Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from mouse MCFD2
Epitope Specificity	1-100/145
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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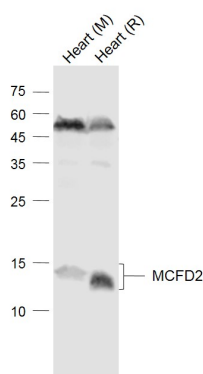
Storage

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.

Background

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Images



Sample:

Lane 1: Heart (Mouse) Lysate at 40 ug

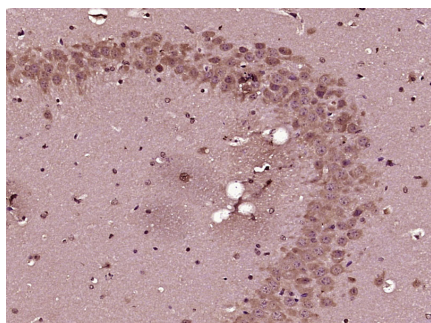
Lane 2: Heart (Rat) Lysate at 40 ug

Primary: Anti-MCFD2 (AP94135) at 1/1000 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 16 kD

Observed band size: 14 kD



Paraformaldehyde-fixed, paraffin embedded (Mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (MCFD2) Polyclonal Antibody, Unconjugated (AP94135) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.

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