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ABCB11 Recombinant Rabbit mAb

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Catalog # AP94283

Product Information

Application	WB, IHC-P, IHC-F, IF
Reactivity	Human
Predicted	Mouse
Host	Rabbit
Clonality	Recombinant
Calculated MW	146 KDa
Physical State	Liquid
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	Membrane; Multi-pass membrane protein.
SIMILARITY	Belongs to the ABC transporter superfamily. ABCB family. Multidrug resistance exporter (TC 3.A.1.201) subfamily. Contains 2 ABC transmembrane type-1 domains. Contains 2 ABC transporter domains.
SUBUNIT	Interacts with HAX1.
DISEASE	Defects in ABCB11 are the cause of progressive familial intrahepatic cholestasis type 2 (PFIC2) [MIM:601847]. PFIC2 is an inherited liver disease of childhood which is characterized by cholestasis and normal serum gamma-glutamyltransferase activity. Defects in ABCB11 are also found in cases of chronic intrahepatic cholestasis without obvious familial history of chronic liver disease. Defects in ABCB11 are the cause of benign recurrent intrahepatic cholestasis type 2 (BRIC2) [MIM:605479]. BRIC is characterized by intermittent episodes of cholestasis without progression to liver failure. There is initial elevation of serum bile acids, followed by cholestatic jaundice which generally spontaneously resolves after periods of weeks to months. The cholestatic attacks vary in severity and duration and patients are asymptomatic between episodes, both clinically and biochemically.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The membrane-associated protein encoded by this gene is a member of the superfamily of ATP-binding cassette (ABC) transporters. ABC proteins transport various molecules across extra- and intra-cellular membranes. ABC genes are divided into seven distinct subfamilies (ABC1, MDR/TAP, MRP, ALD, OABP, GCN20, White). This protein is a member of the MDR/TAP subfamily. Members of the MDR/TAP subfamily are involved in multidrug resistance. The protein encoded by this gene is the major canalicular bile salt export pump in man. Mutations in this gene cause a form of progressive familial intrahepatic cholestases which are a group of inherited disorders with severe cholestatic liver disease from early infancy. [provided by RefSeq, Jul 2008]

Additional Information

Target/Specificity	Expressed predominantly, if not exclusively in the liver, where it was further localized to the canalicular microvilli and to subcanalicular vesicles of the hepatocytes by in situ.
Dilution	WB=1:500-1000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.

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