

# ABCB11 Recombinant Rabbit mAb

ABCB11 Recombinant Rabbit mAb Catalog # AP94283

#### **Product Information**

**Application** WB, IHC-P, IHC-F, IF

Host Rabbit
Clonality Recombinant
Calculated MW 146 KDa
Physical State Liquid
Isotype IgG

**Purity** affinity purified by Protein A

Buffer

SUBCELLULAR LOCATION

SIMILARITY

0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.

Membrane; Multi-pass membrane protein.

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=1:500-1:1000,IHC-P=1:100-500,IHC-F=,IF=0

Format 0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce

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

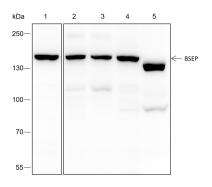
reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody

is stable for at least two weeks at 2-4 °C.

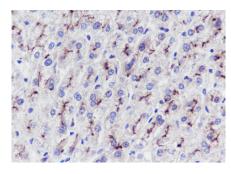
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## **Images**



Blocking buffer: 5% NFDM/TBST Primary ab dilution: 1:1000 Primary ab incubation condition: 2 hours at room temperature Secondary ab: Goat Anti-Rabbit IgG H&L (HRP) Lysate: 1: Jurkat, 2: HepG2, 3: A549, 4: LNCaP, 5: Mouse liver Protein loading quantity: 20 µg Exposure time: 60 s Predicted MW: 146 kDa Observed MW: 146 kDa



Tissue: Human liver Section type: Formalin fixed & Paraffin -embedded section Retrieval method: High temperature and high pressure Retrieval buffer: Tris/EDTA buffer, pH 9.0 Primary ab dilution: 1:500 Primary ab incubation condition: 1 hour at room temperature Secondary ab: SP Kit(Rabbit) (sp-0023) Counter stain: Hematoxylin (Blue) Comment: Color brown is the positive signal for AP94283

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