

bs-1954R-A647

• Rabbit Anti-BSEP/ABCB11 Polyclonal Antibody, Alexa Fluor 647 conjugated

Conjugated Primary Antibodies

Background:

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 transporter in humans and mice. Mutations in the human gene cause a form of progressive familial intrahepatic cholestases which are a group of inherited disorders with severe cholestatic liver disease from early infancy.

Bile formation is an important function of the liver. It is mediated by hepatocytes which generate bile flow within the bile canaliculi by continuous vectorial secretion of bile salts and other solutes across their canalicular (apical) membrane. Bile secretion is mediated by several ATP-binding cassette (ABC) transporters located in the canalicular membrane of hepatocytes. Among these ABC transporters, ABCB11 (bile salt export pump or BSEP) represents the primary, if not sole transport system for the canalicular excretion of bile salts. Bile secretory failure results in cholestasis and progressive familial intrahepatic cholestasis (PFIC) in infancy represents a group of inherited cholestatic diseases that are classified into three subtypes. One of these subtypes, PFIC II, is associated with mutations in the ABCB11 gene. PFIC patients with mutations in the ABCB11 gene have normal serum gamma-glutamyltransferase activity, low concentrations of bile salts in bile, and an absence of bile duct proliferation. Additionally, human obesity is associated with altered cholesterol homeostasis including increased production and turnover, as well as secretion of excess cholesterol from the liver into bile.

ABCB11 is a multifunctional polypeptide with two homologous halves, each containing a hydrophobic membrane anchoring domain and an ATP binding cassette (ABC) domain. The membrane anchoring domain is composed of six helices buried in the lipid bilayer of the plasma membrane and the ATP binding ABCs are exposed to the cytosol. The membrane anchoring domain helices are thought to form channels spanning the plasma membrane.

Purification: Was purified by Protein A and peptide affinity chromatography.

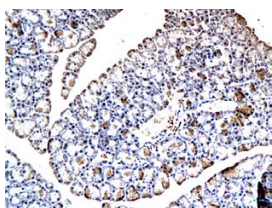
Storage:

Prepared as lyophilized powder or liquid and shipped on ice. Store at -20°C for one year. Protect from light.

Reconstitution:

If the antibody is in liquid form, no reconstitution needed.

Reconstitution is only required for the lyophilized antibody. Please refer to the reconstitution instruction card in the package.



Size: 100ul or 100ug lyophilized

Concentration: 1ug/uL

Host: Rabbit

Reactivities: Human, Mouse, Rat, Rabbit,

Application:

- IF (1:100-500)
- Not yet tested in other applications. Optimal working dilutions must be determined by the end user.

Antibody Type: Polyclonal

Isotype: IgG

Molecular Weight: 145kDa

Preservatives:

10ug/uL BSA and 0.1% NaN₃.

For research use only. CAUTION: Not for human or animal therapeutic or diagnostic use.

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