

**Rabbit Anti-ABCB4/MDR3 Polyclonal: RC0009, RC0009RTU7**

**Intended Use:** For Research Use Only

**Description:** Mediates ATP-dependent export of organic anions and drugs from the cytoplasm. Hydrolyzes ATP with low efficiency. Human MDR3 is not capable of conferring drug resistance. Mediates the translocation of phosphatidylcholine across the canalicular membrane of the hepatocyte. Defects in ABCB4 are the cause of progressive familial intrahepatic cholestasis type 3 (PFIC3). PFIC3 is an autosomal recessive liver disorder presenting with early onset cholestasis that progresses to cirrhosis and liver failure before adulthood. It is characterized by elevated serum gamma-glutamyltransferase levels. Defects in ABCB4 are a cause of intrahepatic cholestasis of pregnancy (ICP); also known as obstetric cholestasis. ICP is a multifactorial liver disorder of pregnancy. It presents during the second or, more commonly, the third trimester of pregnancy with intense pruritus which becomes more severe with advancing gestation and cholestasis. Cholestasis results from abnormal biliary transport from the liver into the small intestine. ICP causes fetal distress, spontaneous premature delivery and intrauterine death. ICP patients have spontaneous and progressive disappearance of cholestasis after delivery. Defects in ABCB4 are a cause of gallbladder disease type 1 (GBD1). It is one of the major digestive diseases. Gallstones composed of cholesterol (cholelithiasis) are the common manifestations in western countries. Most people with gallstones, however, remain asymptomatic through their lifetimes.

**Specifications**

Clone:	Polyclonal
Source:	Rabbit
Isotype:	IgG
Reactivity:	Human, mouse
Localization:	Membrane
Formulation:	Antibody in PBS pH7.4, containing BSA, Proclin 300, glycerol, < 0.05% sodium azide (NaN <sub>3</sub> )
Storage:	Store at 2°- 8°C.
Applications:	IHC, ELISA, WB
Package:	

Description	Catalog No.	Size
ABCB4/MDR3 Polyclonal Concentrated	RC0009	1 ml
ABCB4/MDR3 Polyclonal Prediluted	RC0009RTU7	7 ml

**IHC Procedure\***

Positive Control Tissue:	HCC, heart tissue lysates
Concentrated Dilution:	10-50
Pretreatment:	Citrate pH6.0 or EDTA pH8.0, 15 minutes using Pressure Cooker, or 30-60 minutes using water bath at 95°-99°C
Incubation Time and Temp:	30-60 minutes @ RT
Detection:	Refer to the detection system manual

\* Result should be confirmed by an established diagnostic procedure.

**References:**

1. Functional analysis of ABCB4 mutations relates clinical outcomes of progressive familial intrahepatic cholestasis type 3 to the degree of MDR3 floppase activity. Gordo-Gilart R, et al. Gut. Jan;64(1):147-55, 2015.
2. LPAC syndrome associated with deletion of the full exon 4 in a ABCB4 genetic mutation in a patient with hepatitis C. Fombuena B, et al. Rev Esp Enferm Dig. 2014 Dec;106(8):544-7, 2014.
3. Aspects of liver pathology in adult patients with MDR3/ABCB4 gene mutations. Wendum D, et al. Virchows Arch. Mar;460(3):291-8, 2012.
4. ABCB4 heterozygous gene mutations associated with fibrosing cholestatic liver disease in adults. Zioli M, et al. Gastroenterology. Jul;135(1):131-41, 2008.
5. Dietary lecithin protects against cholestatic liver disease in cholic acid-fed Abcb4- deficient mice. Lamireau T, et al. Pediatr Res. Feb;61(2):185-90, 2007.