Medaysis
Enable Innovation

DATA SHEET

Mouse Anti-ATP8B1/PFIC1 Polyclonal: RC0008-1

Intended Use: For Research Use Only

Description: Found in most tissues except brain and skeletal muscle. Most abundant in pancreas and small intestine. May play a role in the transport of aminophospholipids from the outer to the inner leaflet of various membranes and the maintenance of asymmetric distribution of phospholipids in the canicular membrane. May have a role in transport of bile acids into the canaliculus, uptake of bile acids from intestinal contents into intestinal mucosa or both. Defects in ATP8B1 are the cause of progressive familial intrahepatic cholestasis type 1 (PFIC1); also known as Byler disease. PFIC1 is an autosomal recessive disorder, characterized by early infancy cholestasis, that may be initially episodic but progresses to malnutrition, growth retardation and end-stage liver disease before adulthood. Defects in ATP8B1 are the cause of benign recurrent intrahepatic cholestasis type 1 (BRIC1); also known as Summerskill syndrome. 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. Patients are asymptomatic between episodes, both clinically and biochemically.

Specifications

Clone:	Polyclonal
Source:	Rabbit
Isotype:	IgG
Reactivity:	Human
Localization:	Membrane, cytoplasm
Formulation:	Purified antibody in PBS pH7.4, containing Glycerol and < 0.05% sodium azide (NaN3)
Storage:	Store at 2°- 8°C.
Applications:	IHC, IF, WB

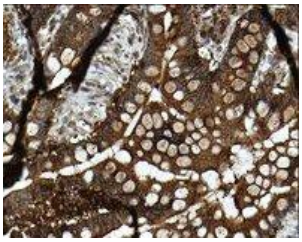
Package:

Description	Catalog No.	Size
ATP8B1/PFIC1 Polyclonal Concentrated	RC0008-1	0.1 ml

IHC Procedure*

Positive Control Tissue:	Skeletal muscle
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.



Human colon FFPE tissue stained with anti-ATP8B1 using DAB

References:

1. Identification of ATP8B1 as a blood-brain barrier-enriched protein. Haas MJ, et al. Cell Mol Neurobiol. May;34(4):473-8, 2014.
2. ATP8B1 deficiency in mice reduces resistance of the canalicular membrane to hydrophobic bile salts and impairs bile salt transport. Paulusma CC, et al. Hepatology. Jul;44(1):195-204, 2006.
3. FIC1, the protein affected in two forms of hereditary cholestasis, is localized in the cholangiocyte and the canalicular membrane of the hepatocyte. Eppens EF, et al. J Hepatol. Oct;35(4):436-43, 2001.

Orders: customercare@medaysis.com Support: techsupport@medaysis.com Tel: 888-608-3167 www.medaysis.com