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ABCA3/P180 Lamellar Body Protein Rabbit pAb

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Catalog # AP56753

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q99758
Predicted	Rat, Dog, Human, Mouse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	191362
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human ABCA3/P180 Lamellar Body Protein
Epitope Specificity	65-160/1704
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.
DISEASE	Defects in ABCA3 are the cause of pulmonary surfactant metabolism dysfunction type 3 (SMDP3) [MIM:610921]; also called pulmonary alveolar proteinosis due to ABCA3 deficiency. A rare lung disorder due to impaired surfactant homeostasis. It is characterized by alveolar filling with floccular material that stains positive using the periodic acid-Schiff method and is derived from surfactant phospholipids and protein components. Excessive lipoproteins accumulation in the alveoli results in severe respiratory distress.
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 intracellular membranes. ABC genes are divided into seven distinct subfamilies (ABC1, MDR/TAP, MRP, ALD, OABP, GCN20, White). This protein is a member of the ABC1 subfamily. Members of the ABC1 subfamily comprise the only major ABC subfamily found exclusively in multicellular eukaryotes. The full transporter encoded by this gene may be involved in development of resistance to xenobiotics and engulfment during programmed cell death. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	21
Other Names	Phospholipid-transporting ATPase ABCA3, 7.6.2.1, ABC-C transporter, ATP-binding cassette sub-family A member 3, ATP-binding cassette transporter 3, ATP-binding cassette 3, Xenobiotic-transporting ATPase ABCA3, 7.6.2.2, 150 Kda mature form, ABCA3 (HGNC:33), ABC3

Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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.

Protein Information

Name	ABCA3 (HGNC:33)
Synonyms	ABC3
Function	Catalyzes the ATP-dependent transport of phospholipids such as phosphatidylcholine and phosphoglycerol from the cytoplasm into the lumen side of lamellar bodies, in turn participates in the lamellar bodies biogenesis and homeostasis of pulmonary surfactant (PubMed:16959783, PubMed:17574245, PubMed:27177387, PubMed:28887056, PubMed:31473345). Transports preferentially phosphatidylcholine containing short acyl chains (PubMed:27177387). In addition plays a role as an efflux transporter of miltefosine across macrophage membranes and free cholesterol (FC) through intralumenal vesicles by removing FC from the cell as a component of surfactant and protects cells from free cholesterol toxicity (PubMed:25817392, PubMed:26903515, PubMed:27177387).
Cellular Location	Endosome, multivesicular body membrane; Multi-pass membrane protein. Cytoplasmic vesicle membrane. Late endosome membrane. Lysosome membrane Note=Localized in the limiting membrane of lamellar bodies in lung alveolar type II cells (PubMed:11718719, PubMed:16959783, PubMed:22673903, PubMed:24142515, PubMed:27177387). Trafficks via the Golgi, sorting vesicles (SVs) and late endosome/multivesicular body network directly to the outer membrane of lamellar bodies in AT2 lung epithelial cells or to lysosomes and lysosomal-related organelles (LROs) in other cells where undergoes proteolytic cleavage and oligosaccharide processing from high mannose type to complex type (PubMed:16959783, PubMed:20863830, PubMed:24142515, PubMed:27177387) Oligomers formation takes place in a post-endoplasmic reticulum compartment (PubMed:27352740).
Tissue Location	Expressed in brain, pancreas, skeletal muscle and heart (PubMed:8706931). Highly expressed in the lung in an AT2-cell- specific manner (PubMed:11718719, PubMed:8706931). Weakly expressed in placenta, kidney and liver (PubMed:8706931). Also expressed in medullary thyroid carcinoma cells (MTC) and in C-cell carcinoma (PubMed:8706931).

Background

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