

SFTPC Recombinant Rabbit mAb

Catalog # AP99162

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P11686
Other Accession	P11686
Host	Rabbit
Clonality	Recombinant
Calculated MW	21013
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human SFTPC
Epitope Specificity	1-50/197
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 BRICHOS domain.
DISEASE	Defects in SFTPC are the cause of pulmonary surfactant metabolism dysfunction type 2 (SMDP2) [MIM:610913]; also called pulmonary alveolar proteinosis due to surfactant protein C deficiency. A rare disease associated with progressive respiratory insufficiency and lung disease with a variable clinical course, 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. Genetic variations in SFTPC are a cause of susceptibility to respiratory distress syndrome in premature infants (RDS) [MIM:267450]; also known as RDS in prematurity. RDS is a lung disease affecting usually premature newborn infants. It is characterized
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes the pulmonary-associated surfactant protein C (SPC), an extremely hydrophobic surfactant protein essential for lung function and homeostasis after birth. Pulmonary surfactant is a surface-active lipoprotein complex composed of 90% lipids and 10% proteins which include plasma proteins and apolipoproteins SPA, SPB, SPC and SPD. The surfactant is secreted by the alveolar cells of the lung and maintains the stability of pulmonary tissue by reducing the surface tension of fluids that coat the lung. Multiple mutations in this gene have been identified, which cause pulmonary surfactant metabolism dysfunction type 2, also called pulmonary alveolar proteinosis due to surfactant protein C deficiency, and are associated with interstitial lung disease in older infants, children, and adults. Alternatively spliced transcript variants encoding different protein isoforms have been identified.

Additional Information

Gene ID	6440
Other Names	BRICD6; PSP-C; SFTP2; SMDP2; SP-C; SP5; SPC; Sftp-2; pro-SpC; PSPC_HUMAN; SFTPC; Pulmonary surfactant-associated protein C; Pulmonary surfactant-associated proteolipid SPL(Val); PSPC_MOUSE; surfactant protein C; surfactant, pulmonary-associated protein C; BRICHOS domain containing 6
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,Flow-Cyt=1:50-100,ICC/IF=1:100-200
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	SFTPC (HGNC:10802)
Synonyms	SFTP2
Function	Pulmonary surfactant associated proteins promote alveolar stability by lowering the surface tension at the air-liquid interface in the peripheral air spaces.
Cellular Location	Secreted, extracellular space, surface film.

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