

Rabbit Anti-SP-C antibody

SL10067R

Product Name:	SP-C
Chinese Name:	—————————————————————————————————————
Alias:	PSP C; PSPC; Pulmonary surfactant apoprotein 2; pulmonary surfactant apoprotein PSP C; pulmonary surfactant associated protein C; pulmonary surfactant associated proteolipid SPL pVal; Pulmonary surfactant associated proteolipid SPL(Val); SFTP 2; SFTP2; SFTPC; SFTPC surfactant pulmonary associated protein C; SP 5; SP C; SP5; SPC; surfactant associated protein pulmonary 2; Surfactant protein c; Surfactant pulmonary associated protein C; PSPC_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Rabbit,Sheep,
Applications:	ELISA=1:500-1000Flow-Cyt=1µg/TestIF=1:100-500 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	4/21kDa
Cellular localization:	Extracellular matrixSecretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human SP-C:24-58/197
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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.

Function:

Pulmonary surfactant associated proteins promote alveolar stability by lowering the surface tension at the air-liquid interface in the peripheral air spaces.

Subcellular Location:

Secreted, extracellular space, surface film.

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 by deficient gas exchange, diffuse atelectasis, high-permeability lung edema and fibrin-rich alveolar deposits called 'hyaline membranes'.

Similarity:

Contains 1 BRICHOS domain.

SWISS: P11686

Gene ID: 6440

Database links:

Entrez Gene: 6440Human

Entrez Gene: 20389 Mouse

Omim: 178620Human

