10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

CFTR C-terminus Recombinant Mouse mAb

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Catalog # AP94229

Product Information

Application	IHC-P, IHC-F, IF
Host	Rabbit
Clonality	Recombinant
Physical State	Liquid
Isotype	IgG2a, Kappa
Purity	affinity purified by Protein G
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Early endosome membrane; Multi-pass membrane protein. Cell membrane.
SIMILARITY	Belongs to the ABC transporter superfamily. ABCC family. CFTR transporter (TC 3.A.1.202) subfamily. Contains 2 ABC transmembrane type-1 domains. Contains 2 ABC transporter domains.
SUBUNIT	Interacts with SHANK2 (By similarity). Interacts with SLC9A3R1, MYO6 and GOPC. Interacts with SLC4A7 through SLC9A3R1. Found in a complex with MYO5B and RAB11A. Interacts with ANO1.
Post-translational modifications	Phosphorylated; activates the channel. It is not clear whether PKC phosphorylation itself activates the channel or permits activation by phosphorylation at PKA sites. Phosphorylated by AMPK. Ubiquitinated, leading to its degradation in the lysosome. Deubiquitination by USP10 in early endosomes, enhances its endocytic recycling.
DISEASE	Cystic fibrosis (CF) [MIM:219700]: A common generalized disorder of the exocrine glands which impairs clearance of secretions in a variety of organs. It is characterized by the triad of chronic bronchopulmonary disease (with recurrent respiratory infections), pancreatic insufficiency (which leads to malabsorption and growth retardation) and elevated sweat electrolytes. It is the most common genetic disease in Caucasians, with a prevalence of about 1 in 2'000 live births. Inheritance is autosomal recessive. Note=The disease is caused by mutations affecting the gene represented in this entry. Congenital bilateral absence of the vas deferens (CBAVD) [MIM:277180]: Important cause of sterility in men and could represent an incomplete form of cystic fibrosis, as the majority of men suffering from cystic fibrosis lack the vas deferens. Note=The disease is caused by mutations affecting the gene represented in this entry.
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 a member of the ATP-binding cassette (ABC) transporter superfamily. ABC proteins transport various molecules across extra- and intra-cellular membranes. ABC genes are divided into seven distinct subfamilies (ABC1, MDR/TAP, MRP, ALD, OABP, GCN20, White). This protein is a member of the MRP subfamily that is involved in multi-drug resistance. The encoded protein functions as a chloride channel and controls the regulation of other transport pathways. Mutations in this gene are associated with the autosomal recessive disorders cystic fibrosis and congenital bilateral aplasia of the vas deferens. Alternatively spliced transcript variants have been

described, many of which result from mutations in this gene. [provided by RefSeq, Jul 2008]

Additional Information

Target/Specificity	Found on the surface of the epithelial cells that line the lungs and other organs.
Dilution	IHC-P=1: 50-1: 200,IHC-F=,IF=0
Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
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.

Background

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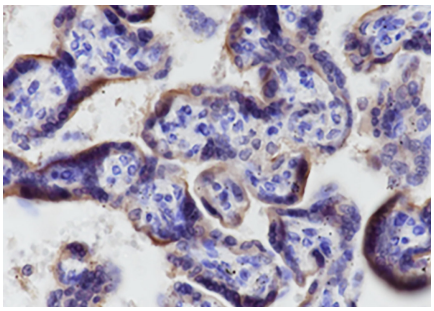
Images



Tissue: Human placenta Section type: Formalin fixed & Paraffin -embedded section Retrieval method: High temperature and high pressure Retrieval buffer: Tris/EDTA buffer, pH 9.0 Primary ab dilution: 1:100 Primary ab incubation condition: 1 hour at room temperature Secondary ab: SP Kit(Mouse)(sp-0024) Counter stain: Hematoxylin (Blue) Comment: Color brown is the positive signal for AP94229