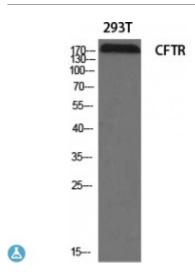


Anti-CFTR antibody



Description Rabbit polyclonal to CFTR.

Model STJ92253

Host Rabbit

Reactivity Human, Mouse, Rat

Applications ELISA, IHC, WB

Immunogen Synthesized peptide derived from human CFTR around the non-

phosphorylation site of S737.

Immunogen Region 680-760 aa

Gene ID 1080

Gene Symbol CFTR

Dilution range WB 1:500-1:2000IHC 1:100-1:300ELISA 1:5000

Specificity CFTR Polyclonal Antibody detects endogenous levels of CFTR protein.

Tissue Specificity Expressed in the respiratory airway, including bronchial epithelium, and in the

female reproductive tract, including oviduct (at protein level) . Detected in pancreatic intercalated ducts in the exocrine tissue, on epithelial cells in intralobular striated ducts in sublingual salivary glands, on apical membranes of crypt cells throughout the small and large intestine, and on the reabsorptive duct in skin sweat glands . Detected on the equatorial segment of the sperm

head (at protein level) . Detected in na

Purification The antibody was affinity-purified from rabbit antiserum by affinity-

chromatography using epitope-specific immunogen.

Note For Research Use Only (RUO).

Protein Name Cystic fibrosis transmembrane conductance regulator CFTR ATP-binding

cassette sub-family C member 7 Channel conductance-controlling ATPase

cAMP-dependent chloride channel

Molecular Weight 168 kDa

Clonality Polyclonal

Conjugation Unconjugated

Isotype IgG

Formulation Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Concentration 1 mg/ml

Storage Instruction Store at -20°C, and avoid repeat freeze-thaw cycles.

Database Links HGNC:1884OMIM:219700

Alternative Names Cystic fibrosis transmembrane conductance regulator CFTR ATP-binding

cassette sub-family C member 7 Channel conductance-controlling ATPase

cAMP-dependent chloride channel

Function Epithelial ion channel that plays an important role in the regulation of

epithelial ion and water transport and fluid homeostasis. Mediates the transport of chloride ions across the cell membrane. Channel activity is coupled to ATP hydrolysis. The ion channel is also permeable to HCO(3-); selectivity depends on the extracellular chloride concentration. Exerts its function also by modulating the activity of other ion channels and transporters . Plays an important role in airway fluid homeostasis . Contributes to the regulation of the pH and the ion content of the airway surface fluid layer and thereby plays an important role in defense against pathogens. Modulates the activity of the epithelial sodium channel (ENaC) complex, in part by regulating the cell surface expression of the ENaC complex . Inhibits the activity of the ENaC channel containing subunits SCNN1A, SCNN1B and SCNN1G. Inhibits the activity of the ENaC channel containing subunits SCNN1D, SCNN1B and SCNN1G, but not of the ENaC channel containing subunits SCNN1A, SCNN1B and SCNN1G. May regulate bicarbonate secretion and salvage in epithelial cells by regulating the transporter SLC4A7 . Can inhibit the chloride channel activity of ANO1 . Plays a role in the

and capacitation.

Sequence and Domain Family Binds and hydrolyzes ATP via the two cytoplasmic ABC transporter

nucleotide-binding domains . The two ATP-binding domains interact with each other, forming a head-to-tail dimer . Normal ATPase activity requires interaction between the two domains . The first ABC transporter nucleotide-binding domain has no ATPase activity by itself . The PDZ-binding motif mediates interactions with GOPC and with the SLC4A7, SLC9A3R1/EBP50 complex. The R region is intrinsically disordered . It mediates channel activation when it is phosphorylated, but not in the absence of

chloride and bicarbonate homeostasis during sperm epididymal maturation

phosphorylation.

Cellular Localization Apical cell membrane Early endosome membrane Cell membrane Recycling

endosome membrane Endoplasmic reticulum membrane. The channel is internalized from the cell surface into an endosomal recycling compartment,

Post-translational Modifications

from where it is recycled to the cell membrane . In the oviduct and bronchus, detected on the apical side of epithelial cells, but not associated with cilia .

N-glycosylated. Phosphorylated; cAMP treatment promotes phosphorylation and activates the channel . Dephosphorylation decreases the ATPase activity (in vitro) . Phosphorylation at PKA sites activates the channel .

Phosphorylation at PKC sites enhances the response to phosphorylation by

 $\ensuremath{\mathsf{PKA}}$. Phosphorylated by $\ensuremath{\mathsf{AMPK}}$; this inhibits channel activity .

Ubiquitinated, leading to its degradation in the lysosome . Deubiquitination by USP10 in early endosomes enhances its endocytic recycling to the cell

membrane . Ubiquitinated by RNF185 during ER stress .

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