

**#2269** Store at  $-20^{\circ}\text{C}$

# CFTR Antibody

100  $\mu\text{l}$   
 (10 Western mini-blot)



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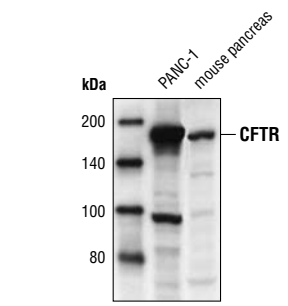
This product is for *in vitro* research use only and is not intended for use in humans or animals.  
 This product is not intended for use as a therapeutic or in diagnostic procedures.

Applications	Species Cross-Reactivity*	Molecular Wt.	Source
W	H, M, R, Mk	168 kDa	Rabbit**

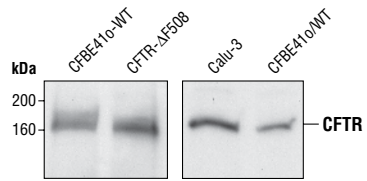
**Background:** CFTR (ABC35, ABC7, CBAVD, CF, dj760C5.1, MRP7, TNR-CFTR) is a member of the ATP-binding cassette (ABC) transporter superfamily. Mutations in ABC genes have been linked to many diseases. CFTR is a plasma membrane cyclic AMP activated chloride channel that is expressed in the epithelial cells of the lung and several other organs (1,2). It mediates the secretion of  $\text{Cl}^-$  and also regulates several channels including the epithelial sodium channel (ENaC),  $\text{K}^+$  channels, ATP release mechanisms, anion exchangers, sodium bicarbonate transporters and aquaporin water channels (3,4,5,6,7,8,9,10). Mutations in the CFTR gene cause cystic fibrosis, a disease that is characterized by exocrine pancreatic insufficiency, increase in sweat gland  $\text{NaCl}$ , male infertility and airway disease (1,2,11). Intracellular trafficking regulates the number of CFTR molecules at the cell surface, which in part regulates  $\text{Cl}^-$  secretion. Deletion of phenylalanine 508 (deltaF508) is the most common mutation in CF patients. This mutation results in retention in the ER, where ER quality control mechanisms target the deltaF508 mutant to the proteasome for degradation (12–15). Therefore, disruption of CFTR trafficking leads to dysregulation of  $\text{Cl}^-$  secretion at the plasma membrane of epithelial cells.

**Specificity/Sensitivity:** CFTR Antibody detects endogenous levels of total CFTR protein.  
**Source/Purification:** Polyclonal antibodies are produced by immunizing animals with a synthetic peptide (KLH-coupled) corresponding to amino acids near the amino terminus of human CFTR. Antibodies are purified by protein A and peptide affinity chromatography.

**Background References:**  
 (1) Bradbury, N.A. et al. (1999) *Am. J. Physiol.* 276, L659–L668.  
 (2) Bertrand, C.A. and Frizzell, R.A. (2003) *Am. J. Physiol. Cell Physiol.* 285, C1–C18.  
 (3) Ko, S.B. et al. (2004) *Nat. Cell Biol.* 6, 343–350.  
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 (6) Stutts, M.J. et al. (1997) *J. Biol. Chem.* 272, 14037–14040.



Western blot analysis of extracts from PANC-1 cells and whole mouse pancreas using CFTR Antibody.



Western blot analysis of extracts of human airway epithelial cells, Calu-3 (expressing endogenous CFTR) and CFBE41o (stably expressing WT-CFTR or deltaF508-CFTR), using CFTR Antibody. Kindly provided by Dr. Agnes Swiatecka-Urban and Dr. Bruce A. Stanton, Department of Physiology, Dartmouth Medical School, Hanover, NH.

(7) Cheung, K.H. et al. (2003) *Biol. Reprod.* 68, 1505–15010.  
 (8) Shumaker, H. et al. (1999) *Am. J. Physiol.* 276, C16–C25.  
 (9) Schwiebert, E.M. et al. (1999) *Physiol. Rev.* 79, S145–S166.  
 (10) Yoo, D. et al. (2004) *J. Biol. Chem.* 279, 6863–6873.  
 (11) Cohn, J.A. et al. (2005) *Hum. Mutat.* 26, 303–307.  
 (12) Gibson, R.L. et al. (2003) *Am. J. Respir. Crit. Care Med.* 168, 918–951.  
 (13) Boucher, R.C. (2004) *Eur. Respir. J.* 23, 146–158.  
 (14) Riordan, J.R. (2005) *Annu. Rev. Physiol.* 67, 701–718.

**IMPORTANT: For Western blots, incubate membrane with diluted antibody in 5% w/v BSA, 1X TBS, 0.1% Tween-20 at 4°C with gentle shaking, overnight.**

**Storage:** Supplied in 10 mM sodium HEPES (pH 7.5), 150 mM NaCl, 100  $\mu\text{g}/\text{ml}$  BSA and 50% glycerol. Store at  $-20^{\circ}\text{C}$ . Do not aliquot the antibody.

**\*Species cross-reactivity is determined by Western blot.**  
**\*\*Anti-rabbit secondary antibodies must be used to detect this antibody.**

**Recommended Antibody Dilutions:**  
 Western blotting 1:1000

**For application specific protocols please see the web page for this product at [www.cellsignal.com](http://www.cellsignal.com).**

**Companion Products:**  
 Phototope<sup>®</sup>-HRP Western Blot Detection System, Anti-rabbit IgG, HRP-linked Antibody #7071  
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 Prestained Protein Marker, Broad Range (Premixed Format) #7720  
 Biotinylated Protein Ladder Detection Pack #7727  
 20X LumiGLO<sup>®</sup> Reagent and 20X Peroxide #7003

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