

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human CFTR. Specifically recognizes <i>in vitro</i> synthesized CFTR, recombinant CFTR protein, and non-recombinant CFTR protein (1, 2).
Source	Monoclonal Mouse IgG ₁ Clone # 13-1
Purification	Protein A or G purified from hybridoma culture supernatant
Immunogen	β-galactosidase-coupled CFTR Cys590-Lys830 (predicted) Accession # P13569
Formulation	Supplied as a solution in PBS containing BSA. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 μm filtered solution in PBS.

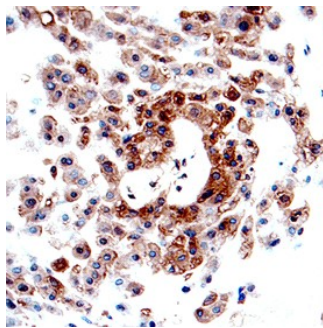
APPLICATIONS

Please Note: Optimal dilutions should be determined by each laboratory for each application. General Protocols are available in the Technical Information section on our website.

	Recommended Concentration	Sample
Western Blot	1 μg/mL	Human CFTR transfected cell line
Immunohistochemistry	8-25 μg/mL	See Below
Immunoprecipitation	1-2 μg/10 ⁶ cells	T84 human colon carcinoma cell line, see our available Western blot detection antibodies
Immunofluorescence	Cheng, S.H. <i>et al.</i> (1990) <i>Cell</i> 63 :827. Marino, C.R. <i>et al.</i> (1991) <i>J. Clin. Invest.</i> 88 :712.	

DATA

Immunohistochemistry



CFTR in Human Placenta. CFTR was detected in immersion fixed paraffin-embedded sections of human placenta using Mouse Anti-Human CFTR R Domain Monoclonal Antibody (Catalog # MAB1660) at 15 μg/mL overnight at 4 °C. Before incubation with the primary antibody tissue was subjected to heat-induced epitope retrieval using Antigen Retrieval Reagent-Basic (Catalog # CTS013). Tissue was stained using the Anti-Mouse HRP-DAB Cell & Tissue Staining Kit (brown; Catalog # CTS002) and counterstained with hematoxylin (blue). Specific labeling was localized to the plasma membrane and cytoplasm of decidual cells. View our protocol for [Chromogenic IHC Staining of Paraffin-embedded Tissue Sections](#).

PREPARATION AND STORAGE

Shipping	The product is shipped with dry ice or equivalent. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
Stability & Storage	Use a manual defrost freezer and avoid repeated freeze-thaw cycles. <ul style="list-style-type: none"> ● 12 months from date of receipt, -20 to -70 °C, as supplied. ● 1 month, 2 to 8 °C under sterile conditions after opening. ● 6 months, -20 to -70 °C under sterile conditions after opening.

BACKGROUND

Cystic fibrosis transmembrane conductance regulator (CFTR) is a multi-pass transmembrane protein that functions as a chloride channel. CFTR belongs to the ATP-binding cassette (ABC) superfamily. Mutations in CFTR cause the pulmonary disease, cystic fibrosis (CF). Specifically, deletion of phenylalanine at position 508 (DeltaF508-CFTR) results in a folding defect which impairs chloride channel function. The mechanism by which channel dysfunction relates to disease symptoms is a focus of intense research. CFTR dysfunction results in disruption of ion transport and subsequent blockage of airways by secreted mucus. CFTR may also play a role in the skeletal muscle atrophy and dysfunction that characterizes CF. In addition, CFTR-mediated chloride secretion underlies fluid accumulation and cyst growth in autosomal dominant polycystic kidney disease (ADPKD).

References:

1. Gregory, R.J. *et al.* (1990) *Nature* **347**:328.
2. Cheng, S.H. *et al.* (1990) *Cell* **63**:827.