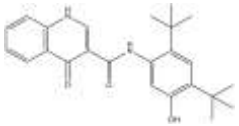
PRODUCT: Ivacaftor

ALTERNATE NAME: N-[2,4-bis(1,1-dimethylethyl)-5-hydroxyphenyl]-1,4-dihydro-4-oxo-3-quinolinecarboxamide; Cystic Fibrosis Transmembrane Conductance Regulator Potentiator; VX770; Kalydeco

CATALOG #: 9582-5, 25

AMOUNT: 5 mg, 25 mg

STRUCTURE:



MOLECULAR FORMULA: $C_{24}H_{28}N_2O_3$

MOLECULAR WEIGHT: 392.49

CAS NUMBER: 873054-44-5

APPEARANCE: Off-white solid

SOLUBILITY: DMSO (>50 mg/ml)

PURITY: ≥98% by HPLC

STORAGE: Store at -20℃. Protect from air and light

DESCRIPTION: Ivacaftor (VX-770) is a potent, cell-permeable small molecule potentiator of cystic fibrosis transmembrane conductance regulator (CFTR). It increases the forskolin-induced CFTR-mediated epithelial current in cells expressing the G551D mutation by ~4-fold (EC_{50} = 100 nM) but has no effect on current in the absence of forskolin. Ivacaftor increases chloride secretion in cultured human CF bronchial epithelial cells carrying the G551D mutation on one allele and the common F508del processing mutation on the other allele. It binds CFTR directly and causes CFTR channel opening by an ATP-independent mechanism. It effectively improves sweat chloride *in vivo* while improving lung function when at least one G551D CFTR mutation is present.

REFERENCES:

Van Goor, F., *et al.* (2009). *Proc. Natl. Acad. Sci. USA* **106,** 18825-18830.

HANDLING: Do not take internally. Wear gloves and mask when handling the product! Avoid contact by all modes of exposure.

RELATED PRODUCTS:
CFTR Inhibitor-172 **(2487)**
CFTR Inhibitor, GlyH-101 **(2863)**
PTC-124 **(9421)**
VX-809 **(2857)**

USAGE: ***FOR RESEARCH CH USE ONLY! Not to be used in humans***

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