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Data Sheet

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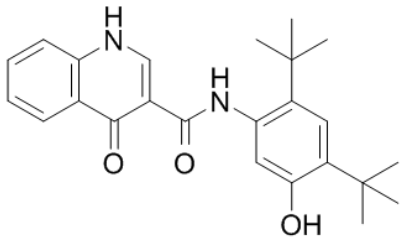
Product Name	:	Ivacaftor
Cat. No.	:	PC-43325
CAS No.	:	873054-44-5
Molecular Formula	:	$C_{24}H_{28}N_2O_3$
Molecular Weight	:	392.4907
Target	:	CFTR
Solubility	:	10 mM in DMSO



Biological Activity

Ivacaftor (VX-770, VX770) is a potent, orally bioavailable **CFTR potentiator**, increases G551D- and F508del CFTR-mediated Cl- secretion with EC50 of 100 nM.
Ivacaftor (VX-770) increases CFTR channel open probability (Po) in both the F508del processing mutation and the G551D gating mutation in recombinant cells, increases the Po of G551D CFTR by 6-fold, the Po of F508del- and wild-type CFTR by 5-fold and 2-fold, respectively.
Ivacaftor (VX-770) also potentiates CFTR-mediated Cl− secretion in primary cultures of G551D/F508del HBE and F508del HBE.
Ivacaftor (VX-770) demonstrates potential for treatment of cystic fibrosis with certain mutations in CFTR gene (primarily the G551D mutation).

References

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Accurso FJ, et al. ***N Engl J Med.*** 2010 Nov 18;363(21):1991-2003.

Yu W, et al. ***J Biol Chem.*** 2011 Jul 15;286(28):24714-25.

Hadida S, et al. ***J Med Chem.*** 2014 Dec 11;57(23):9776-95.

Caution: Product has not been fully validated for medical applications. Lab Use Only!

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