

## Ivacaftor benzenesulfonate

## Chemical Properties

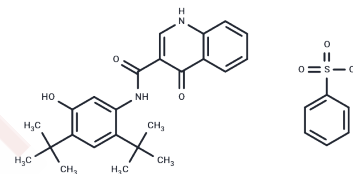
CAS No. : 1134822-09-5

Formula: C<sub>30</sub>H<sub>34</sub>N<sub>2</sub>O<sub>6</sub>S

Molecular Weight: 550.67

Storage: Powder: -20°C for 3 years | In solvent: -80°C for 1 year

Actual storage temperature shall be subject to the COA.



## Biological Description

Description	Ivacaftor benzenesulfonate, used for cystic fibrosis treatment, is an orally bioavailable CFTR potentiator.
Targets(IC50)	Others,CFTR,Autophagy
In vitro	Ivacaftor, at a concentration of 10 $\mu$ M, significantly enhances CFTR activity in cells expressing the W1282X mutation, showing superior efficacy compared to cells with the R1162X mutation. It exhibits negligible activity towards 160 other targets, including the GABAA benzodiazepine receptor. Additionally, ivacaftor notably enhances chloride secretion, demonstrating a 10-fold increase in potency with an EC <sub>50</sub> of 0.236 $\pm$ 0.200 $\mu$ M compared to F508del HBEs. In recombinant cells, VX-770 (ivacaftor) markedly improves the CFTR channel's open probability (P <sub>o</sub> ) in the presence of both F508del processing and G551D gating mutations, and amplifies forskolin-stimulated IT in temperature-corrected F508del-FRT cells approximately 6-fold at an EC <sub>50</sub> of 25 nM. Ivacaftor triples PC secretion activity for the ABCB4-G535D mutation and significantly amplifies it for ABCB4-G536R (13.7-fold), ABCB4-S1076C (6.7-fold), ABCB4-S1176L (9.4-fold), and ABCB4-G1178S (5.7-fold), effectively correcting the functional defects of these ABCB4 mutations.
In vivo	Ivacaftor (1-200 mg/kg, p.o.) demonstrates high oral bioavailability in rats.

### Preparing Stock Solutions

	1mg	5mg	10mg
1 mM	1.816 mL	9.0798 mL	18.1597 mL
5 mM	0.3632 mL	1.816 mL	3.6319 mL
10 mM	0.1816 mL	0.908 mL	1.816 mL
50 mM	0.0363 mL	0.1816 mL	0.3632 mL

Please select the appropriate solvent to prepare the stock solution, according to the solubility of the product in different solvents. Please use it as soon as possible.

Note: The dilution table applies only to solid products. For liquid products, please calculate the stock solution based on the stated concentration and/or density.

### Reference

Delaunay JL, et al. Functional defect of variants in the adenosine triphosphate-binding sites of ABCB4 and their rescue by the cystic fibrosis transmembrane conductance regulator potentiator, ivacaftor (VX-770). *Hepatology*. 2017 Feb;65(2):560-570

Mutyam V, et al. Therapeutic benefit observed with the CFTR potentiator, ivacaftor, in a CF patient homozygous for the W1282X CFTR nonsense mutation. *J Cyst Fibros*. 2017 Jan;16(1):24-29

Hadida S, et al. Discovery of N-(2,4-di-tert-butyl-5-hydroxyphenyl)-4-oxo-1,4-dihydroquinoline-3-carboxamide (VX-770, ivacaftor), a potent and orally bioavailable CFTR potentiator. *J Med Chem*. 2014 Dec 11;57(23):9776-9

Van Goor F, et al. Rescue of CF airway epithelial cell function in vitro by a CFTR potentiator, VX-770. *Proc Natl Acad Sci U S A*. 2009 Nov 3;106(44):18825-30.

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