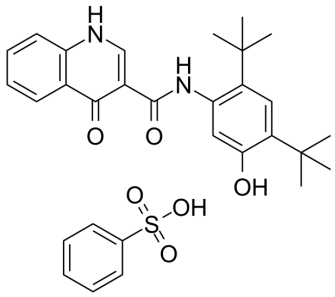
Ivacaftor benzenesulfonate

Cat. No.:	HY-13017A
CAS No.:	1134822-09-5
Molecular Formula:	$C_{30}H_{34}N_2O_6S$
Molecular Weight:	550.67
Target:	CFTR; Autophagy
Pathway:	Membrane Transporter/Ion Channel; Autophagy
Storage:	Please store the product under the recommended conditions in the Certificate of Analysis.



BIOLOGICAL ACTIVITY

Description	Ivacaftor benzenesulfonate is an orally bioavailable CFTR potentiator, used for cystic fibrosis treatment.
In Vitro	Ivacaftor (10 μM) increases the PC secretion activity by 3-fold for ABCB4-G535D, 13.7-fold for ABCB4-G536R, 6.7-fold for ABCB4-S1076C, 9.4-fold for ABCB4-S1176L, and 5.7-fold for ABCB4-G1178S. Ivacaftor corrects the functional defect of ABCB4 mutants[1]. Ivacaftor (10 μM) significantly increases CFTR activity in W1282X-expressing cells compared to R1162X CFTR cells [2]. Ivacaftor shows no significant activity against 160 targets tested including the $GABA_A$ benzodiazepine receptor. Ivacaftor increases the chloride secretion with an EC_{50} of 0.236 ± 0.200 μM, a 10-fold shift in potency compared to the F508del HBEs[3]. In recombinant cells, VX-770 increases CFTR channel open probability (Po) in both the F508del processing mutation and the G551D gating mutation. VX-770 increases forskolin-stimulated I_T in temperature-corrected F508del-FRT cells by appr 6-fold with an EC_{50} of 25 nM[4]. MCE has not independently confirmed the accuracy of these methods. They are for reference only.
In Vivo	Ivacaftor (1-200 mg/kg, p.o.) exhibits good oral bioavailability in rat[3]. MCE has not independently confirmed the accuracy of these methods. They are for reference only.

CUSTOMER VALIDATION

- Front Cell Dev Biol. 2021 May 11;9:678209.
- J Cell Sci. 2022 Jan 21;jcs.259002.
- Org Process Res Dev. 2019, 23, 11, 2302-2322.

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REFERENCES

[1]. 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

[2]. 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

[3]. 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

[4]. 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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