Ivacaftor hydrate

Cat. No.: HY-13017B

CAS No.: 1134822-07-3

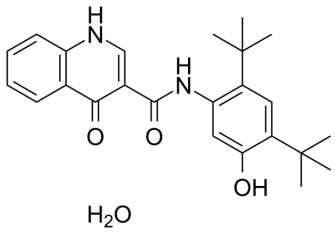
Molecular Formula: $C_{24}H_{30}N_2O_4$

Molecular Weight: 410.51

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 hydrate (VX-770 hydrate) 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.

Caution: Product has not been fully validated for medical applications. For research use only.

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