



Ivacaftor (benzenesulfonate)

Catalog No: tcsc1595

Available Sizes
Size: 5mg
Size: 10mg
Size: 50mg
Size: 100mg
Specifications
CAS No: 1134822-09-5
Formula: $C_{30}^{H}_{34}^{N}_{2}^{O}_{6}^{S}$
Pathway: Membrane Transporter/Ion Channel
Target: CFTR
Purity / Grade: >98%
Solubility: 10 mM in DMSO
Alternative Names: VX-770 benzenesulfonate
Observed Molecular Weight: 550.67



Product Description

Ivacaftor benzenesulfonate is an orally bioavailable **CFTR** potentiator, used for cystic fibrosis treatment.

In Vitro: Ivacaftor (10 μ M) 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 μ M) 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 EC₅₀ of 0.236 \pm 0.200 μ M, 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 EC₅₀ of 25 nM^[4].

In Vivo: Ivacaftor (1-200 mg/kg, p.o.) exhibits good oral bioavailability in rat^[3].

All products are for RESEARCH USE ONLY. Not for diagnostic & therapeutic purposes!