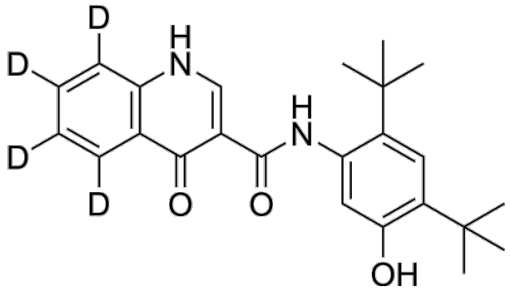
Product Data Sheet

Product Name: Ivacaftor-d4
Cat. No.: GC70634

Chemical Properties

Cas. No.			
Formula	$C_{24}H_{24}D_4N_2O_3$	M.Wt	396.52
Solubility	DMSO : 50 mg/mL (126.10 mM; Need ultrasonic; Hygroscopic DMSO has a significant impact on the solubility of product, please use newly opened DMSO)	Storage	4°C, sealed storage, away from moisture
General tips	For obtaining a higher solubility , please warm the tube at 37 °C and shake it in the ultrasonic bath for a while.Stock solution can be stored below -20°C for several months.		
Shipping Condition	Evaluation sample solution : ship with blue ice All other available size: ship with RT , or blue ice upon request.		
Structure			



Background

Ivacaftor-d4 (VX-770-d4) is the deuterium labeled-Ivacaftor . Ivacaftor is a potent and orally active CFTR potentiator, targeting G551D-CFTR and F508del-CFTR with EC50s of 100 nM and 25 nM, respectively.

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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} value of 0.236 ± 0.200 μM, a 10-fold shift in potency compared to the F508del HBEs[3].
In recombinant cells, Ivacaftor increases CFTR channel open probability (Po) in both the F508del processing mutation and the G551D gating mutation. Ivacaftor increases forskolin-stimulated I_T in temperature-corrected F508del-FRT cells by appr 6-fold with an EC_{50} of 25 nM[4].

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