

Product Data Sheet

Product Name: Ivacaftor hydrate

Cat. No.: GC13316

Chemical Properties

Cas. No. 1134822-07-3

Chemical Name (Z)-N-(2,4-di-tert-butyl-5-hydroxyphenyl)-4-oxo-1,4-dihydroquinoline-3-carbimidic acid hydrate

SMILES CC(C)(C1=CC(C(C)(C)C)=C(O)C=C1/N=C(O)/C2=CNC3=CC=CC=C3C2=O)C.O

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

M.Wt 410.51

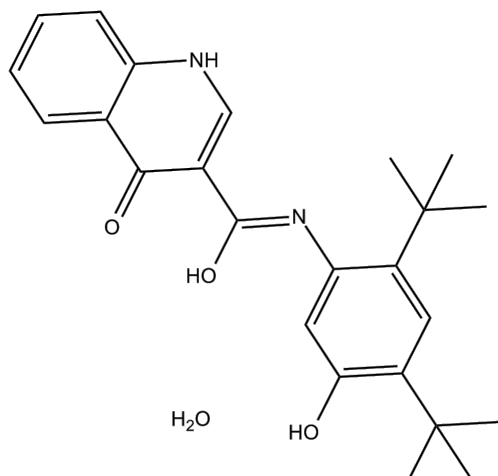
Solubility Soluble in DMSO

Storage Store at -20°C

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

Description: IC₅₀ Value: 25 nM (F508del-CFTR); 100 nM (G551D-CFTR) [1] Ivacaftor (VX-770) is a CFTR Potentiator approved for patients with the G551D mutation of cystic fibrosis, which accounts for 4-5% cases of cystic fibrosis. In vitro: In recombinant cells VX-770 increased CFTR channel open probability (P_o) in both the F508del processing mutation and the G551D gating mutation. VX-770 also increased Cl⁻ secretion in cultured human CF bronchial epithelia (HBE) carrying the G551D

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

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gating mutation on one allele and the F508del processing mutation on the other allele by approximately 10-fold, to approximately 50% of that observed in HBE isolated from individuals without CF [1]. in vivo: At day 28, in the group of subjects who received 150 mg of VX-770, the median change in the nasal potential difference (in response to the administration of a chloride-free isoproterenol solution) from baseline was -3.5 mV (range, -8.3 to 0.5; $P=0.02$ for the within-subject comparison, $P=0.13$ vs. placebo), and the median change in the level of sweat chloride was -59.5 mmol per liter (range, -66.0 to -19.0; $P=0.008$ within-subject, $P=0.02$ vs. placebo) [2]. Toxicity: Six severe adverse events occurred in two subjects (diffuse macular rash in one subject and five incidents of elevated blood and urine glucose levels in one subject with diabetes). All severe adverse events resolved without the discontinuation of VX-770 [2]. Clinical trial: Roll-Over Study of Ivacaftor in Cystic Fibrosis Pediatric Subjects With a CFTR Gating Mutation. Phase 3

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