

## Product Data Sheet

Product Name: Ivacaftor benzenesulfonate

Cat. No.: GC12489

### Chemical Properties

Cas. No. 1134822-09-5

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

SMILES CC(C)(C1=CC(C(C)(C)C)=C1)C(=O)C=C1/N=C(O)/C2=CNC3=CC=CC=C3C2=O)C.OS(C4=CC=CC=C4)(=O)=O

Formula  $C_{30}H_{34}N_2O_6S$  M.Wt 550.67

Solubility  $\geq 55.1$  mg/mL in DMSO,  $\geq 13.86$  mg/mL in EtOH with gentle warming 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 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

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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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