



Product Specification Sheet

Product Name: VX-770 (Ivacaftor)

Catalog Number: C8770

Technical information:

Chemical Formula: $C_{24}H_{28}N_2O_3$

CAS #: 873054-44-5

Molecular Weight: 392.49

Purity: > 98%

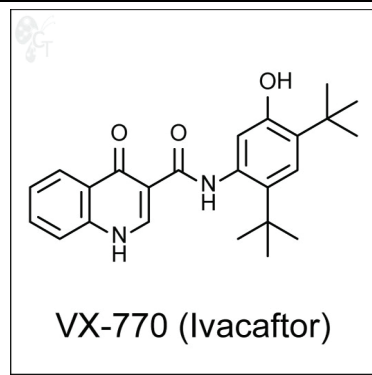
Appearance: Off-white solid

Solubility: Soluble in DMSO up to 100 mM

Chemical Name: N-(2,4-di-tert-butyl-5-hydroxyphenyl)-4-oxo-1,4-dihydroquinoline-3-carboxamide

Storage: For longer shelf life, store solid powder or DMSO solution at -20°C desiccated.

Shelf Life: In the unopened package, powder is stable for 1 year and DMSO solution is stable for 6 months under proper storage condition.



- Handling:**
- To make 10 mM stock solution, add 0.255mL of DMSO for each mg of VX-770 (Ivacaftor).
 - For DMSO solution, briefly spin the vial at 500 rpm in a 50 mL conical tube to ensure maximum sample recovery.

Biological Activity: VX-770 (Ivacaftor) is a CFTR potentiator and has been shown to potentiate normal CFTR, as well as CFTR with G551D and F508-del mutations. Ivacaftor directly binds to the ion channel, causes conformation change and open the ion channel to improve chloride transportation. In primary cultured human CF bronchial epithelia (HBE) carrying the G551D and F508del CFTR mutations, Ivacaftor (10 μ M) potently increases the forskolin-stimulated Cl⁻ secretion with an EC50 of 236 nM. [1]

VX-770 (Ivacaftor) is approved for cystic fibrosis patients with G551D mutation, and has shown efficacy in a patient with S549N mutation. [2]

- Reference:**
1. 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; 106(44):18825-30. Pubmed ID: 19846789
 2. McGarry ME and Nielson DW. Normalization of sweat chloride concentration and clinical improvement with ivacaftor in a patient with cystic fibrosis with mutation S549N. Chest. 2013; 144(4):1376-8. Pubmed ID: 24081349

To reorder: <http://www.cellagentech.com/VX-770-Ivacaftor/>

For Technical Support: technical@cellagentech.com

Chemicals are sold for research use only, not for clinical or diagnostic use.