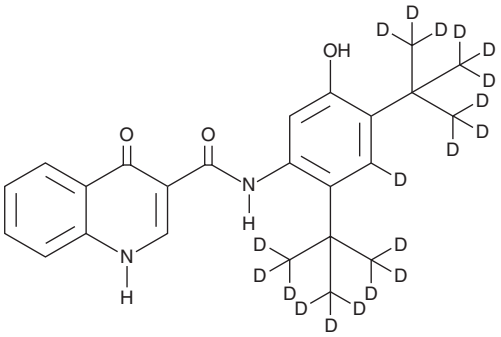
# PRODUCT INFORMATION

## Ivacaftor-$d_{19}$

*Item No. 28539*

| | |
|---|---|
| **CAS Registry No.:** | 1413431-22-7 |
| **Formal Name:** | N-[2,4-*bis*[1,1-di(methyl-$d_3$)ethyl-2,2,2-$d_3$]-5-hydroxyphenyl-3d]-1,4-dihydro-4-oxo-3-quinolinecarboxamide |
| **MF:** | $C_{24}H_9D_{19}N_2O_3$ |
| **FW:** | 411.6 |
| **Chemical Purity:** | ≥98% (Ivacaftor) |
| **Deuterium Incorporation:** | ≥99% deuterated forms ($d_1$-$d_{19}$); ≤1% $d_0$ |
| **Supplied as:** | A solid |
| **Storage:** | -20°C |
| **Stability:** | ≥4 years |



*Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.*

### Laboratory Procedures

Ivacaftor-$d_{19}$ is intended for use as an internal standard for the quantification of ivacaftor (Item No. 15145) by GC- or LC-MS. The accuracy of the sample weight in this vial is between 5% over and 2% under the amount shown on the vial. If better precision is required, the deuterated standard should be quantitated against a more precisely weighed unlabeled standard by constructing a standard curve of peak intensity ratios (deuterated *versus* unlabeled).

Ivacaftor-$d_{19}$ is supplied as a solid. A stock solution may be made by dissolving the ivacaftor-$d_{19}$ in the solvent of choice, which should be purged with an inert gas. Ivacaftor-$d_{19}$ is soluble in organic solvent DMSO. Ivacaftor-$d_{19}$ is also soluble in a 1:1 solution of acetonitrile:methanol.

### Description

Ivacaftor is an orally bioavailable potentiator of the cystic fibrosis transmembrane conductance regulator (CFTR) that improves chloride transport.[1] It increases the forskolin-induced CFTR-mediated epithelial current in cells expressing the G551D missense mutation associated with severe cystic fibrosis by approximately 4-fold ($EC_{50}$ = 100 nM) but has no effect on current in the absence of forskolin.[1] Ivacaftor increases chloride secretion in cultured human cystic fibrosis bronchial epithelial cells carrying the G551D mutation on one allele and the common ΔF508 processing mutation on the other allele.[1] It binds CFTR directly and leads to CFTR channel opening *via* an ATP-independent mechanism.[2] Formulations containing ivacaftor have been used in the treatment of cystic fibrosis in patients carrying one or more mutations in the *CFTR* gene.

### References


1. Van Goor, F., Hadida, S., Grootenhuis, P.D., *et al.* Rescue of CF airway epithelial cell function in vitro by a CFTR potentiator, VX-770. Proc. *Natl. Acad. Sci. USA* **106(44)**, 18825-18830 (2009).
2. Eckford, P.D., Li, C., Ramjeesingh, M., et al. Cystic fibrosis transmembrane conductance regulator (CFTR) potentiator VX-770 (ivacaftor) opens the defective channel gate of mutant CFTR in a phosphorylation-dependent but ATP-independent manner. *J. Biol. Chem.* **287(44)**, 36639-36649 (2012).





**CAYMAN CHEMICAL**
1180 EAST ELLSWORTH RD
ANN ARBOR, MI 48108 · USA
**PHONE:** [800] 364-9897
[734] 971-3335
**FAX:** [734] 971-3640
CUSTSERV@CAYMANCHEM.COM
WWW.**CAYMANCHEM**.COM