

Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

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PRODUCT INFORMATION



Ivacaftor-d₁₉ Item No. 28539

CAS Registry No.: 1413431-22-7

Formal Name: N-[2,4-bis[1,1-di(methyl-d₂)ethyl-2,2,2-

d₂]-5-hydroxyphenyl-3d]-1,4-dihydro-4-

oxo-3-quinolinecarboxamide

MF: $C_{24}H_9D_{19}N_2O_3$

411.6 FW:

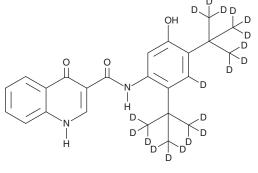
Chemical Purity: ≥98% (Ivacaftor)

Deuterium

 \geq 99% deuterated forms (d₁-d₁₉); \leq 1% d₀ Incorporation:

Supplied as: A solid -20°C Storage: Stability: ≥2 years

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



Laboratory Procedures

lvacaftor-d₁₉ 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-d₁₉ is supplied as a solid. A stock solution may be made by dissolving the ivacaftor-d₁₉ in the solvent of choice. Ivacaftor-d₁₉ is soluble in organic solvent DMSO, which should be purged with an inert gas. Ivacaftor- d_{10} 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. 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 (EC₅₀ = 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 Δ F508 processing mutation on the other allele. 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).

WARNING
THIS PRODUCT IS FOR RESEARCH ONLY - NOT FOR HUMAN OR VETERINARY DIAGNOSTIC OR THERAPEUTIC USE.

This material should be considered hazardous until further information becomes available. Do not ingest, inhale, get in eyes, on skin, or on clothing. Wash thoroughly after handling. Before use, the user must review the complete Safety Data Sheet, which has been sent via email to your institution.

WARRANTY AND LIMITATION OF REMEDY

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