

Additive Potentiation of R334W-CFTR Function by Novel Small Molecules

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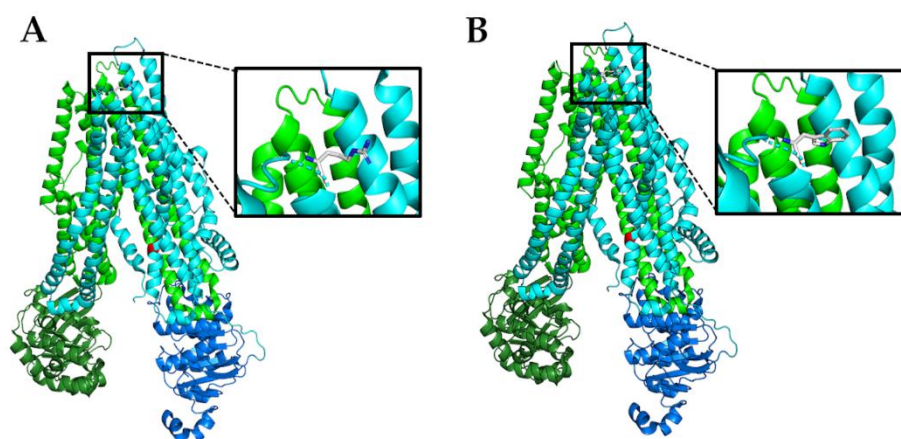


Figure S1. Ribbon diagram of the dephosphorylated, ATP-free human CFTR structure (PDB: 5UAK) demonstrating the location of the R334 residue in WT-CFTR (**A**) and mutant R334W-CFTR (**B**).

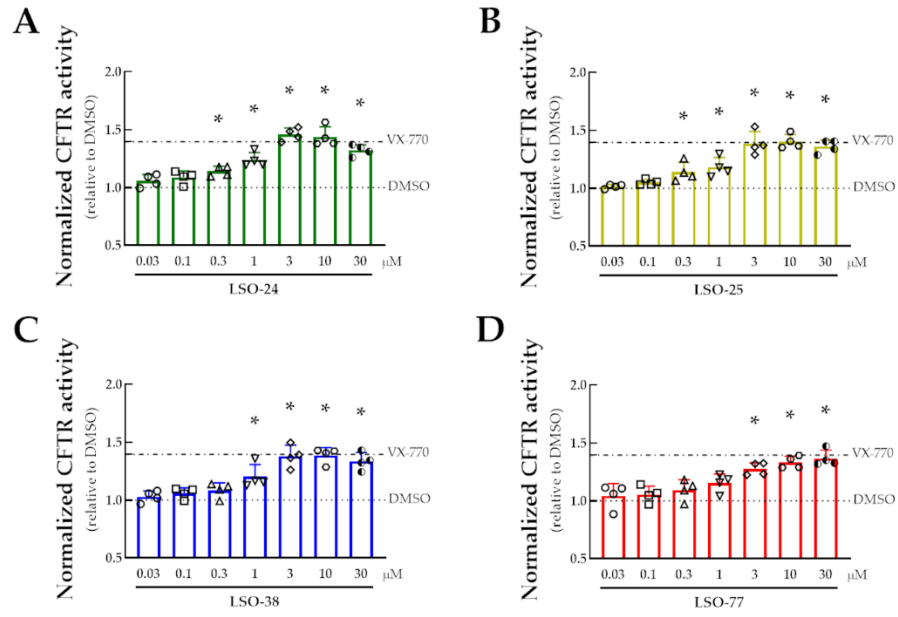


Figure S2. Dose-response relationships for the compounds (A) LSO-24, (B) LSO-25, (C) LSO-38 and (D) LSO-77 were determined by the HS-YFP assay on a plate reader in CFBE cells expressing R334W-CFTR. *P < 0.05 *vs.* DMSO.

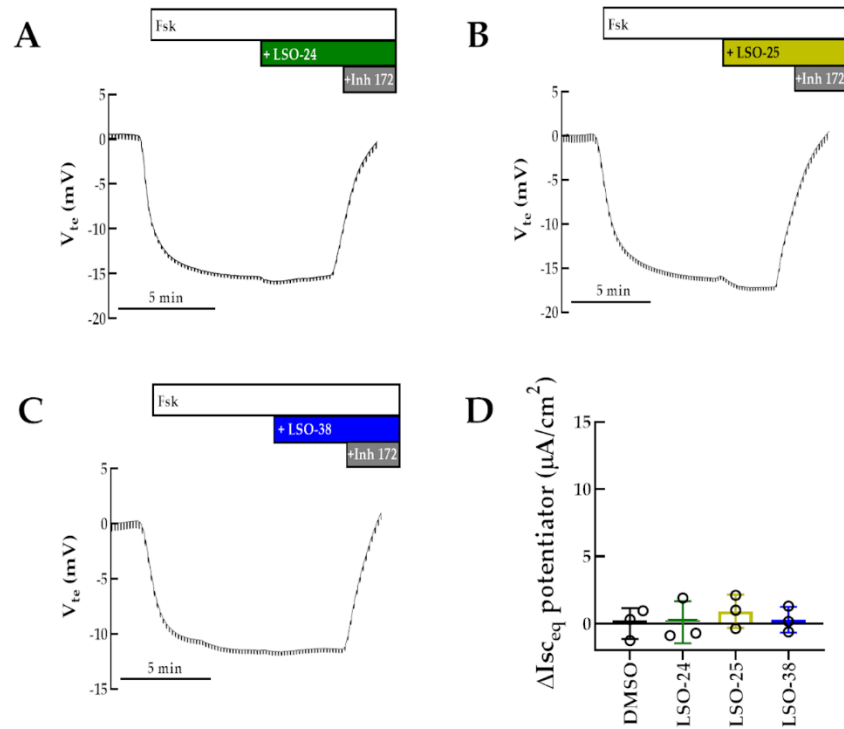


Figure S3. (A–C) Representative Ussing chamber (open-circuit) recording representing transepithelial voltage measurements (V_{te}) of polarized monolayers of CFBE cells expressing WT-CFTR. Cells were acutely subsequently stimulated with Fsk (0.128 μM), test compounds (DMSO, 5 μM LSO-24, 5 μM LSO-25 or 5 μM LSO-38) and CFTRInh-172 (30 μM). (D) Data are represented as mean (\pm SD) increase in I_{eq} promoted by test potentiator.