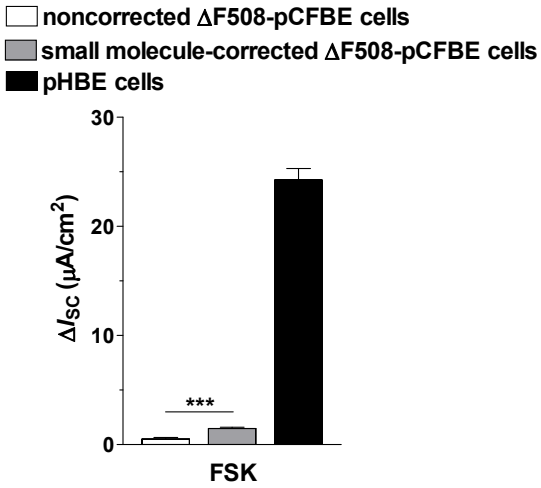
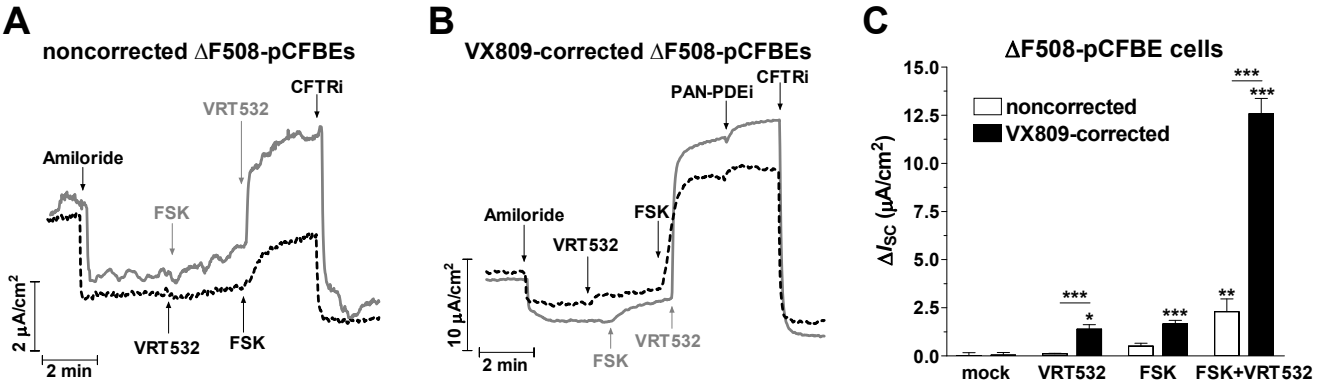
Data Supplement:



Supplementary Figure 1. Comparison of CFTR currents in pHBE and ΔF508-pCFBE cells. Shown is the maximal I_{SC} induced by Forskolin (FSK; 10 μM) in noncorrected or small-molecule-corrected ΔF508-pCFBE cells (combination of experiments with VRT640, VRT325 and VX809 shown in Fig. 4) compared to that of non-CF pHBE cells. Data represent the mean ± sem (n≥6). ***(p<0.001)



Supplementary Figure 2. CFTR correctors, activators and potentiators synergize to maximize CFTR currents in ΔF508-pCFBE cells. Cultures were incubated for 48 h in the presence or absence of the CFTR corrector VX809 (10 μM, basolateral) and CFTR-dependent I_{SC} was then recorded in response to treatment with the CFTR potentiator VRT532 (20 μM) and/or Forskolin (FSK; 10 μM) and the PAN-selective PDE inhibitor IBMX (200 μM) followed by treatment with $CFTR_{inh}$-172 (CFTRi; 20 μM). *A/B*) Representative I_{SC} traces induced by FSK and VRT532 in noncorrected (*A*) and VX809-corrected (*B*) ΔF508-pCFBE cells. *C*) Average I_{SC} induced by VRT532 and/or FSK in noncorrected (n≥5) and VX809-corrected (n≥19) ΔF508-pCFBE cells. Data represent the mean ± sem. ***(p<0.001); **(p<0.01); *(p<0.05)