



SUPPLEMENTARY FIG. S4. Fibroblasts from CF patients with the $\Delta F508$ mutation make a mature distal lung epithelium. (A) Immunocytochemistry showing fibroblasts from $\Delta F508$ patients reprogrammed and express pluripotency genes (NANOG, POU5F1, SOX2, and TRA-1-60). (B) $\Delta F508$ lung endoderm does not express markers of the thyroid lineage (PAX9, TG, and PAX8), the forebrain (PAX6), or the more mature airway epithelium (SFTPC, ABCA3, and MUC5AC). (C) Mature distal airway epithelium expressing the $\Delta F508$ CFTR continues to express distal airway epithelial genes while remaining negative for thyroid (PAX8, PAX9, and TG), forebrain markers (PAX6), and proximal airway lineages (FOXP2, SOX2, and SOX17). White bars = 100 μ M. ** $P \leq 0.01$, *** $P \leq 0.001$, **** $P \leq 0.0001$. FL, human foetal lung control; AL, adult lung control; C, undifferentiated hESC control; $\Delta f508$, airway epithelium from CF patients cultured for 25 days; CF, cystic fibrosis.