

Table s1. Differentially expressed genes/ transcripts, CF vs. non-CF epithelia

Gene/ Transcript	Log2FC -- CF vs. non-CF	adjusted p-value
CYP1A1	4.860221137	7.72E-18
DAPL1	-2.200708195	2.05E-17
LOC105369554	2.932874497	3.48E-17
VWCE	2.101586889	4.32E-12
LGR6	-2.945573223	6.02E-12
VIM	2.506405405	1.66E-10
ITM2A	-2.544195614	1.85E-08
PEG3	-2.229576037	2.02E-08
CTSE	6.897415744	6.57E-08
SERPINA1	2.196424475	9.91E-08
SCGB3A2	9.490325133	1.32E-07
FAIM2	-2.295808508	4.88E-07
SFTPA2	7.817858537	6.92E-07
SFTPB	6.330297523	1.04E-06
LGR5	-3.977247046	1.04E-06
GRP	-3.034999055	1.48E-06
LINC00330	-3.11042107	3.70E-06
ISLR	-2.244397082	5.99E-06
GGT5	3.710849416	6.06E-06
SLC13A2	-2.243103249	6.98E-06
FAM181A-AS1	4.485930485	7.38E-06
TMPRSS11E	-2.430736409	9.80E-06
EDN2	-2.09783869	1.09E-05
LOC101928441	-3.101339989	2.35E-05
LOC284240	-2.734632817	2.74E-05
LOC105371115	2.134146839	2.79E-05
NAPSA	4.800879594	3.74E-05
PGC	7.468932533	4.12E-05
ASCL1	-3.138163701	4.37E-05
HPGD	-2.203601436	4.50E-05
ELMO1	2.196005198	5.33E-05
SFTPA1	8.406424561	7.11E-05
SCG2	-2.300842702	0.000100754
SMOC2	-2.49217645	0.000179392
KRT32	-2.062593989	0.000240337
LMO1	-2.990477739	0.000249682
EXOC3L4	2.470999451	0.00025672
LOC100132154	2.326297571	0.00025672
CXCL13	-4.931575343	0.000274237
CACNA1I	3.335249177	0.000301454
LOC100996747	2.251349056	0.000345156
FAM181A	2.659633828	0.000368593

CRTAC1	2.344900626	0.000369364
SLC16A12	2.35569266	0.000379862
ZNF521	-2.041645082	0.000389049
CYSLTR1	-2.068462067	0.000398852
SUSD2	2.339353575	0.000465238
HS3ST4	3.730720578	0.000481074
NTS	-4.472024123	0.000502744
SCN1A	2.767043808	0.000511267
PADI4	-2.930914388	0.000523505
DGKI	-2.266846614	0.000524741
ZNF503-AS1	2.277168182	0.000540815
BAAT	2.76296584	0.000606243
UGT2A2	-2.153754676	0.000679207
LOC105377548	-3.962949943	0.000788072
C4BPA	3.888771775	0.0007953
C6	2.227048327	0.000890328
LOC105371566	2.466419679	0.000898226
COL1A2	-2.183911408	0.000954797
CYGB	2.74246781	0.001243352
NNMT	2.656666475	0.001267615
TREH	2.38727262	0.001296263
DDC	-2.558949913	0.0015125
SLC38A11	-2.440669498	0.001570546
FABP5	-2.029817489	0.001642785
KRT222	-2.108431271	0.001898724
B3GAT1	2.253131851	0.0021121
AQP1	2.254442514	0.002210126
BCL2L2-PABPN1	7.854436569	0.002279277
SFTA1P	2.473220128	0.002450621
LOC105379005	-2.051213553	0.002609877
SCN3A	-2.197424134	0.002624704
SUCNR1	-2.886175285	0.002690972
MMP1	2.428191601	0.00287857
ZSCAN1	-2.496550645	0.002933817
SCGN	-3.320550459	0.003188055
SOX8	-2.489479468	0.003298466
ATP13A4-AS1	3.30884941	0.003395799
CPLX2	-2.381846754	0.003769508
LTF	2.599969949	0.003769508
LOC105379027	4.10667761	0.00389195
TBC1D3D	2.699688938	0.003974013
LOC105378287	2.155249241	0.003977022
THSD7B	2.705513582	0.004041872
SPARCL1	-2.794172926	0.004223917
NELL1	-2.479352293	0.004844343

CPB1	4.271150985	0.00529914
RTL9	-2.137213865	0.00536603
LOC107986397	6.157100985	0.005563841
RLN2	-3.262353915	0.006724672
CALCA	-2.386357082	0.007218698
GPR4	2.077494369	0.007375269
NPR1	5.609781582	0.008013277
TCEAL2	-2.134990795	0.008107127
SFTPC	6.521669547	0.009238933
OR51E1	-2.262532801	0.009330657
CXCL5	2.306713467	0.009331467
CDH12	-2.193224153	0.009514516
KRT13	-2.348271383	0.009523522
LOC107986800	2.138967859	0.009816457
LOC107987343	2.423625833	0.009989076
EIF2S3B	4.709077626	0.011388953
UGT2A1	-2.179220279	0.01150287
CAMK2A	2.319807773	0.01259535
MS4A15	3.421631515	0.012724113
MSMB	2.509137188	0.014879232
PLA2G1B	3.041534353	0.015349585
PLD5	2.087348599	0.015940084
UGT2B4	-2.023995479	0.016826532
SLC22A31	3.033225477	0.017352337
HP	2.828760977	0.017658535
PEBP4	4.969953718	0.018458822
CSF3R	2.504137704	0.018560348
ETDA	2.364635838	0.021801372
MIR4423	2.298640035	0.022632115
NOS2	-2.434907596	0.023265233
TLX1	-2.250432781	0.023392346
LOC105369526	-2.636190793	0.026895428
SERPIND1	2.88999216	0.028008198
FN1	2.347476635	0.028762958
CD200R1	2.004264391	0.0288502
TRIM39-RPP21	6.09655602	0.029221683
KRT14	-2.377066644	0.03015093
CHGA	-2.158504891	0.030710738
UGT1A8	-2.19708264	0.032418182
SCN7A	-2.272093745	0.033562719
LOC105371856	3.297017325	0.035264936
GUCY1B2	2.146312052	0.036640497
ADGRD1	2.457804579	0.037227231
LINC02242	-2.853438345	0.038194524
NPTX1	-2.134479927	0.038337448

NEU4	-2.272129011	0.038807128
TGM7	-2.294538115	0.040830711
ADRA2A	2.452780536	0.043703854
UGT1A7	-3.444455181	0.048613238

Table s2. Baseline clinical characteristics of study subjects (previously reported in ref. 15)

Genotype	Age	Sex	ppFEV1 (L)	Sweat [Cl ⁻] (mEq/L)
G551D/ ΔF508	23	F	79 (2.8)	99.9
G551D/ ΔF508	25	F	72 (2.42)	102.6
G551D/ ΔF508	23	F	79 (2.46)	90.6
G551D/ ΔF508	30	F	40 (1.11)	65.7
G551D/ ΔF508	27	F	51 (1.49)	91.5
G551D/ 3659delC	22	F	39 (1.25)	109
G551D/ ΔF508	33	F	88 (2.54)	105
G551D/ p67L	57	F	34 (0.82)	80.9
G551D/ ΔF508	35	F	77 (2.16)	98.6
G551D/ G551D	29	M	101 (3.99)	110.3
G551D/ ΔF508	30	M	39 (1.6)	108.2
G551D/ R117H	33	M	72 (2.76)	61.9
R117H/ ΔF508	45	M	108 (3.8)	98
R117H/ ΔF508	40	M	96 (3.75)	81
R117H/ ΔF508	41	F	87 (2.44)	61
R117H/ ΔF508	34	F	62 (1.81)	71
R117H/ ΔF508	40	M	75 (3.05)	78
R117H/ ΔF508	52	M	69 (2.39)	90
R117H/ M156R	42	F	43 (1.37)	61
R117H/ M156R	40	M	35 (1.49)	66
R117H/ 2622+1G→A	25	M	75 (3.41)	89
R117H/ ΔF508	46	F	62 (1.9)	80