

Supplemental Table 1: Key subject characteristics and hepatic fat fraction.

Pt	age	gender	CFTR variant 1 (class)	CFTR variant 2 (class)	diabetes status	insulin	modulat or	PDF F
1	39	male	F508del ^a (2)	F508del (2)	CFRD	Yes	Lum/Iva	0
2	17.5	male	F508del (2)	F508del (2)	CFRD	No	Lum/Iva	0
3	29.9	male	F508del (2)	F508del (2)	CFRD	Yes	Lum/Iva	1.5
4	37.5	male	F508del (2)	F508del (2)	CFRD	Yes	Lum/Iva	1.9
5	36.2	male	F508del (2)	F508del (2)	CFRD	Yes	Lum/Iva	2
6	26.4	female	F508del (2)	F508del (2)	CFRD	Yes	Lum/Iva	2
7	18.8	female	F508del (2)	F508del (2)	CFRD	Yes	Lum/Iva	2.3
8	19.6	male	F508del (2)	F508del (2)	NGT	No	Lum/Iva	2.4
9	26.8	male	F508del (2)	G542X ^b (1)	NGT	No	none	2.7
10	35.6	male	F508del (2)	1898+1G>A ^c (1)	CFRD	No	none	2.8
11	30.6	male	F508del (2)	1717+1G>A ^d (1)	NGT	No	none	3.2
12	19.3	female	F508del (2)	3659delC ^e (1)	CFRD	Yes	none	3.6
13	11.3	male	F508del (2)	1717-1G>A ^d (1)	NGT	No	none	3.8
14	22.6	male	I507del ^f (2)	W1282X ^g (1)	CFRD	Yes	none	4.1
15	14.9	male	F508del (2)	R553X ^h (1)	NGT	No	none	6
16	21.9	male	F508del (2)	3659delC ⁱ (1)	NGT	No	none	6.4
17	16.3	male	F508del (2)	F508del (2)	NGT	No	Lum/Iva	6.4
18	36.1	female	F508del (2)	F508del (2)	CFRD	Yes	none	12.5
19	17.0	male	F508del (2)	W1282X ^g (1)	CFRD	Yes	none	14.5
20	13.4	male	F508del (2)	N1303Ki (2)	NGT	No	none	21

CFRD: cystic fibrosis related diabetes; NGT: normal glucose tolerance; Lum/Iva:

Lumacaftor/Ivacaftor ; After each superscript letter is listed the legacy and HVGS 19.01 name for each CFTR variant: ^a F808del, p.Phe508del, c.1521_1523delCTT; ^b G542X, p.Gly542X, c.1624G>T; ^c 1898+1G>A, c.1766+1G->A; ^d 1717+1G->A, c.1585-1G>A; ^e 3659delC, p.Lys1177SerfsX15, c.3528delC; ^f I507del, p.Ile507del, c.1519_1521delATC; ^g W1282X, p.Trp1282X, c.3846G>A; ^h R553X,

p.Arg553X, c.1657C>T; ⁱ 3659delC, p.Lys1177SerfsX15, c.3528delC; ^j N1303K, p.Asn1303Lys, c.3909C>G.
