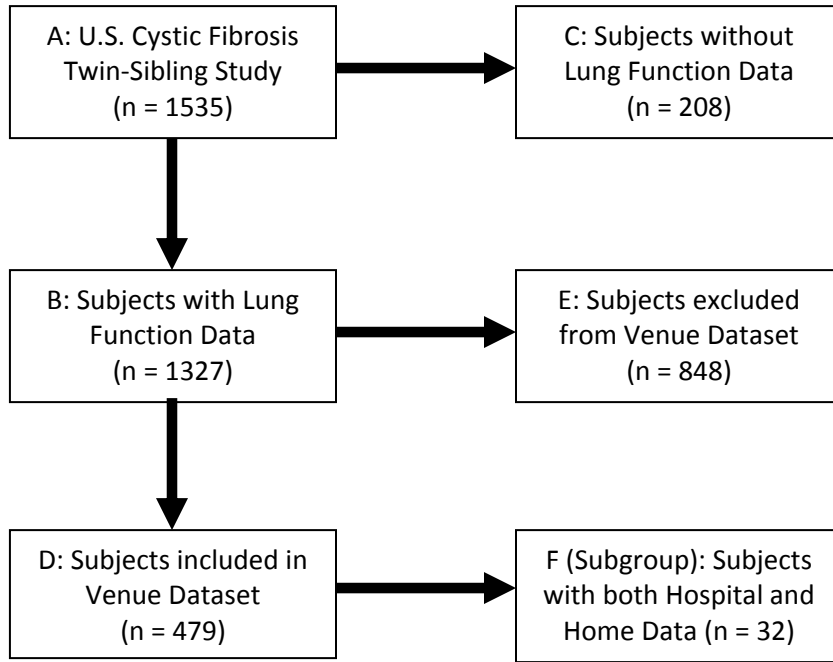


Location and Duration of Treatment of Cystic Fibrosis Respiratory Exacerbations do not Affect Outcomes

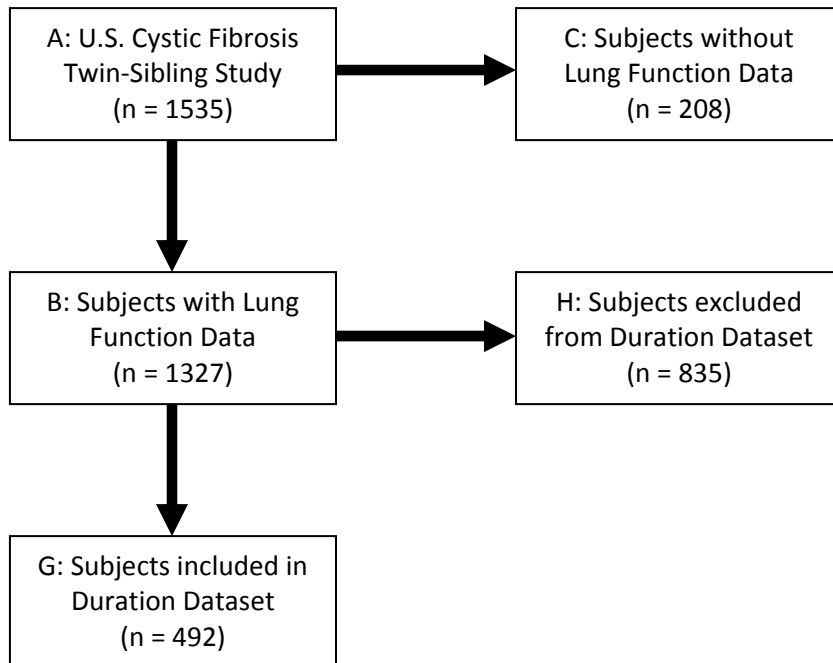
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ONLINE DATA SUPPLEMENT

Supplemental Figure 1. Derivation of Study Population for Venue



Supplemental Figure 2. Derivation of Study Population for Duration



Supplemental Table 1. Demographics of Study Populations

	A Twin- Sibling Study	B Any lung function data ¹	C No lung function data ¹	D Venue of Therapy Group ²	E Excluded from Venue ²	F Subgroup for Hospital vs. Home	G Duration of Therapy Group ³	H Excluded from Duration ³
n	1535	1327	208	479	848	32	492	835
Age (yrs)⁴	16.3 ± 11.6	17.3 ± 9.2	10.3 ± 20.3	19.4 ± 8.3	16.1 ± 9.5	22.1 ± 7.22	19.2 ± 8.4	16.1 ± 9.4
Gender (% Male)	51.8 (n=1534)	52.2	49.8 (n=207)	47.4	54.8	75.0	49.2	53.9
CFTR (% DF508 homozygotes)	49.3 (n=1524)	49.2 (n=1323)	49.8 (n=201)	49.2 (n=478)	49.2 (n=845)	40.6	50.1 (n=491)	48.7 (n=832)

¹ Comparing groups B and C: Age *t* test *p* value<0.0001, gender chi square=0.52, *CFTR* chi square=0.89.

² Comparing groups D and E: Age *t* test *p* value<0.0001, gender chi square=0.009, *CFTR* chi square=0.98.

³ Comparing groups G and H: Age *t* test *p* value<0.0001, gender chi square=0.10, *CFTR* chi square=0.62.

⁴ Age is calculated from the last pulmonary function test used in the study or from 12/31/2007 for subjects without pulmonary function test data.

Supplemental Table 2. Means and Confidence Intervals for Lung Function Measures by Group for Figures 2 and 3

Category	Group	n	Baseline FEV ₁	Pre-therapy FEV ₁	Post-therapy FEV ₁	New Baseline FEV ₁
Venue (Figure 2)	Hospital Only	602	67.4 [65.6, 69.2]	58.8 [57.0, 60.5]	67.9 [66.1, 69.8]	64.1 [62.3, 66.0]
	Home Only	232	65.1 [62.2, 67.9]	59.5 [56.6, 62.4]	64.4 [61.4, 67.5]	61.5 [58.5, 64.6]
	Combination: Hospital & Home	444	71.4 [69.4, 73.3]	63.0 [61.0, 65.0]	72.0 [69.8, 74.1]	67.8 [65.6, 70.0]
Duration (Figure 3)	≤7 days	100	73.6 [68.4, 78.8]	64.3 [59.4, 69.2]	74.4 [69.3, 79.5]	70.9 [65.7, 76.2]
	8-14 days	596	70.4 [69.7, 72.2]	61.6 [59.9, 63.3]	72.0 [70.2, 73.9]	67.6 [65.7, 69.4]
	15-21 days	387	66.3 [64.1, 68.5]	59.1 [56.8, 61.3]	65.7 [63.4, 68.0]	62.6 [60.3, 64.9]
	22-28 days	127	65.7 [62.3, 69.2]	59.5 [55.8, 63.2]	63.7 [59.9, 67.4]	60.2 [56.5, 63.9]
	>28 days	68	59.2 [54.2, 64.1]	52.7 [48.1, 57.4]	57.7 [52.7, 62.8]	55.2 [50.1, 60.2]

Supplemental Table 3. Predictive Models for Baseline Change (n = 832 therapy courses, 342 subjects)

Regression Coefficient (coefficient <i>p</i> value)	Univariate Models	Multivariate Model*	Final Model [†]
Gender (Female = 1, Male = 0)	0.06 (0.93)	0.10 (0.89)	0.05 (0.94)
Number of CFTR F508del alleles (0 - 2)	-0.33 (0.57)	-0.23 (0.67)	
Age at start of therapy (Years)	-0.01 (0.78)	-0.06 (0.23)	-0.05 (0.28)
<i>P. aeruginosa</i> present (Yes = 1; No = 0)	-1.32 (0.42)	-1.70 (0.39)	
<i>B. cepacia</i> complex present (Yes = 1; No = 0)	-0.58 (0.64)	-0.04 (0.97)	
Duration of therapy (Days)	-0.11 (0.02)	-0.15 (0.004)	-0.15 (0.001)
Baseline FEV₁ (Knudson percentage)	-0.03 (0.05)	-0.01 (0.50)	
FEV₁ drop prior to therapy (Sick Decline) (Baseline FEV ₁ – PreFEV ₁)	0.26 (<0.001)	0.27 (<0.001)	0.27 (<0.001)
Location (Home = 1, Hospital = 0)	-0.30 (0.69)	0.08 (0.91)	

*Multivariate Model overall *p* value: <0.0001†Final Model overall *p* value: <0.0001; *r* = 0.36; residual kurtosis: 5.64.**Supplemental Table 4.** Predictive Models for Baseline Change (n = 832 therapy courses, 342 subjects)

Regression Coefficient (coefficient <i>p</i> value)	Univariate Models	Multivariate Model*	Final Model [†]
Gender (Female = 1, Male = 0)	0.06 (0.93)	0.03 (0.96)	0.07 (0.91)
Number of CFTR F508del alleles (0 - 2)	-0.33 (0.57)	-0.11 (0.81)	
Age at start of therapy (Years)	-0.01 (0.78)	-0.04 (0.32)	-0.04 (0.35)
<i>P. aeruginosa</i> present (Yes = 1; No = 0)	-1.32 (0.42)	-1.39 (0.37)	
<i>B. cepacia</i> complex present (Yes = 1; No = 0)	-0.58 (0.64)	-0.61 (0.58)	
Duration of therapy (Days)	-0.11 (0.02)	-0.08 (0.08)	-0.07 (0.07)
Baseline FEV₁ (Knudson percentage)	-0.03 (0.05)	-0.002 (0.91)	
FEV₁ drop prior to therapy (Sick Decline) (Baseline FEV ₁ – PreFEV ₁)	0.26 (<0.001)	0.54 (<0.001)	0.54 (<0.001)
FEV₁ recovery with therapy (Immediate Recovery) (PostFEV ₁ – PreFEV ₁)	0.08 (0.02)	0.38 (<0.001)	0.37 (<0.001)
Location (Home = 1, Hospital = 0)	-0.30 (0.69)	0.32 (0.66)	

*Multivariate Model overall *p* value: <0.0001

†Final Model overall p value: <0.0001; $r = 0.54$; residual kurtosis: 5.19.

Supplemental Table 5. Predictive Models for Immediate Recovery (n = 832 therapy courses, 342 subjects)

Regression Coefficient (coefficient p value)	Univariate Models	Multivariate Model*	Final Model†
Gender (Female = 1, Male = 0)	-0.27 (0.79)	0.17 (0.81)	-0.04 (0.95)
Number of CFTR F508del alleles (0 - 2)	-0.09 (0.91)	-0.32 (0.59)	
Age at start of therapy (Years)	-0.23 (<0.001)	-0.04 (0.30)	-0.03 (0.43)
<i>P. aeruginosa</i> present (Yes = 1; No = 0)	-1.82 (0.59)	-0.80 (0.64)	
<i>B. cepacia</i> complex present (Yes = 1; No = 0)	1.85 (0.35)	1.52 (0.20)	
Duration of therapy (Days)	-0.33 (<0.001)	-0.20 (<0.001)	-0.21 (<0.001)
Baseline FEV₁ (Knudson percentage)	0.08 (<0.001)	-0.02 (0.15)	
FEV₁ drop prior to therapy (Sick Decline) (Baseline FEV ₁ – PreFEV ₁)	-0.73 (<0.001)	-0.72 (<0.001)	-0.72 (<0.001)
Location (Home = 1, Hospital = 0)	-4.23 (<0.001)	-0.63 (0.46)	

*Multivariate Model overall p value: <0.0001

†Final Model overall p value: <0.0001; $r = 0.66$; residual kurtosis: 4.96.

Supplemental Table 6. Predictive Models for Baseline Change (n = 442 combination therapy courses, 247 subjects)

Regression Coefficient (coefficient p value)	Univariate Models	Multivariate Model*	Final Model†
Gender (Female = 1, Male = 0)	-1.06 (0.34)	-0.79 (0.45)	
Number of CFTR F508del alleles (0 - 2)	0.03 (0.97)	-0.08 (0.92)	
Age at start of therapy (Years)	-0.02 (0.76)	0.01 (0.93)	
<i>P. aeruginosa</i> present (Yes = 1; No = 0)	-3.41 (0.08)	-4.11 (0.03)	-4.42 (0.03)
<i>B. cepacia</i> complex present (Yes = 1; No = 0)	-2.26 (0.19)	-1.77 (0.27)	
Duration of therapy (Days)	-0.15 (0.03)	-0.155 (0.03)	-0.18 (0.01)
Baseline FEV₁ (Knudson percentage)	0.01 (0.82)	0.01 (0.59)	
FEV₁ drop prior to therapy (Sick Decline) (Baseline FEV ₁ – PreFEV ₁)	0.28 (<0.001)	0.29 (<0.001)	0.29 (<0.001)
Percentage of time spent in the hospital (Days in Hospital/Total Days of Treatment)	-1.63 (0.46)	-0.71 (0.75)	

*Multivariate Model overall p value: <0.0001

†Final Model overall p value: <0.0001; $r = 0.34$; residual kurtosis: 4.18.