

**Table 1: Characteristics of Study Population, and by Bronchodilator Responder Status**

Characteristic	All tests	First admission only	Non-Responders (226 tests in 99 subjects)	Responders <sup>1</sup> (23 tests in 17 subjects)	p-value <sup>2</sup>
Tests with significant BD response	23/249 (9.2%)	9/102 (8.8%)			
Age (years), median (IQR) (range)	14.3 (11.7, 16.2) (4.6 – 18.0)	13.6 (10.5, 16.0) (4.6, 18.0)	14.1 (11.5, 16.2) (4.6 – 18.0)	15.2 (13.7, 16.6) (5.8 – 17.8)	0.17
Female, n (%)	141/249 (57%)	57/102 (56%)	56/99 (57%)	5/17 (29%)	0.06
BMI centile, median (IQR) (range)	20.8 (6.8, 37.7) (0.00 – 91.1)	27.5 (15.5, 51.9) (0.00, 91.1)	20.3 (6.4, 37.7) (0.00 – 91.1)	25.4 (8.0, 40.6) (1.5 – 81.1)	0.64
Ethnicity					
Caucasian	186/249 (75%)	82/102 (80%)	79/99 (80%)	15/17 (88%)	0.44
Black	13/249 (5%)	3/102 (3%)	3/99 (3%)	1/17 (6%)	
Asian	11/249 (4%)	7/102 (7%)	7/99 (7%)	0/17 (0%)	
Hispanic	9/249 (4%)	1/102 (1%)	1/99 (1%)	1/17 (6%)	
Other/Mixed	17/249 (7%)	4/102 (4%)	4/99 (4%)	0/17 (0%)	
Missing	13/249 (5%)	5/102 (5%)	5/99 (5%)	0/17 (0%)	
CFTR Mutation Class					
I-III	232/249 (93%)	93/102 (91%)	90/99 (91%)	15/17 (88%)	0.66
IV-V	17/249 (7%)	9/102 (9%)	9/99 (9%)	2/17 (12%)	
Inhaled corticosteroid use	104/249 (42%)	39/102 (38%)	95/226 (42%)	9/23 (39%)	0.83
Baseline FEV <sub>1</sub> %, median (IQR) (range)	70.6 (58.1, 84.6) (25.4 – 121.9)	79.1 (67.3, 91.1) (25.4, 121.2)	71.8 (57.9, 84.9) (25.4 – 121.9)	67.7 (58.6, 77.5) (46.4 – 99.8)	0.31
recovery to 90% of baseline at end of treatment	144/235 (61%)	64/95 (67%)	128/213 (60%)	16/22 (73%)	0.36
Microbiology at admission					
<i>P. aeruginosa</i>	87/247 (35%)	37/100 (37%)	83/225 (37%)	4/22 (18%)	0.10
<i>S. maltophilia</i>	24/247 (10%)	10/100 (10%)	22/225 (10%)	2/22 (9%)	0.99
<i>B. cepacia</i> complex	12/247 (5%)	3/100 (3%)	9/225 (4%)	3/22 (14%)	0.08
<i>S. aureus</i>	114/247 (46%)	44/100 (44%)	103/225 (46%)	11/22 (50%)	0.82
<i>H. influenza</i>	23/247 (9%)	10/100 (10%)	23/225 (10%)	0/22 (0%)	0.24
<i>Aspergillus</i> species	57/247 (23%)	22/100 (22%)	52/225 (23%)	5/22 (23%)	0.99
MRSA	20/247 (8%)	7/100 (7%)	17/225 (8%)	3/22 (14%)	0.40
<i>Achromobacter</i> species	26/247 (11%)	6/100 (6%)	25/225 (11%)	1/22 (5%)	0.49

<sup>1</sup>Responders – defined as a 12% and 200 ml or greater increase in FEV<sub>1</sub> after bronchodilator inhalation.

<sup>2</sup>P value calculated for responders compared to non-responders, using Fisher's exact test for categorical variables and Mann-Whitney test for nonparametric variables.

CFTR – cystic fibrosis transmembrane conductance regulator, MRSA – methicillin resistant Staph aureus, BMI – body mass index, IQR – interquartile range, FEV<sub>1</sub> – forced expiratory volume in one second

Table 2: Pulmonary function (FEV<sub>1</sub>) results for the study cohort

Time	n <sup>1</sup>	Median (IQR) FEV <sub>1</sub> % predicted
Baseline	241	70.6 (58.1, 84.6)
Admission	243	54.4 (41.5, 66.9)
Day 4-10, Pre BD	249	62.3 (48.4, 74.7)
Day 4-10, Post BD	249	65.4 (53.0, 78.3)
End of treatment	243	67.1 (53.8, 78.2)

<sup>1</sup>Pulmonary function data were missing for 8 patients at baseline, and for 6 patients and end of treatment

IQR – interquartile range, FEV<sub>1</sub> – forced expiratory volume in one second, BD – bronchodilator