

**Supplement Tables:**

**Supplement Table 1: Pulmonary Function Testing: Individual Patient Results.**

PT	1			2			3			4			5			6		
	BL	Wk16	Δ	BL	Wk16	Δ	BL	Wk16	Δ	BL	Wk16	Δ	BL	Wk16	Δ	BL	Wk16	Δ
<b>FVC %</b>	53	66	<b>13</b>	84	80	<b>-4</b>	87	91	<b>4</b>	63	72	<b>9</b>	85	77	<b>-7</b>	67	75	<b>8</b>
<b>FEV1 %</b>	54	60	<b>6</b>	71	69	<b>-2</b>	86	87	<b>1</b>	67	77	<b>10</b>	87	79	<b>-8</b>	74	88	<b>14</b>
<b>TLC %</b>	72	82	<b>10</b>	118	114	<b>-4</b>	99	106	<b>7</b>	67	76	<b>9</b>	78	76	<b>-2</b>	80	82	<b>2</b>
<b>DLCO%</b>	55	65	<b>10</b>	89	67	<b>-22</b>	78	70	<b>-8</b>	26	27	<b>1</b>	101	96	<b>-5</b>	61	65	<b>4</b>
PT	7			8			9			10			11			MEDIAN		
	BL	Wk16	Δ	BL	Wk16	Δ	BL	Wk16**	Δ	BL	Wk16	Δ	BL	Wk16	Δ	BL	Wk16	Δ
<b>FVC %</b>	80	82	<b>2</b>	92	71	<b>-21</b>	59	58	<b>-1</b>	62	59	<b>-3</b>	98	105	<b>7</b>	<b>80</b>	<b>75</b>	<b>2</b>
<b>FEV1 %</b>	74	85	<b>11</b>	74	76	<b>6</b>	63	58	<b>-5</b>	62	65	<b>3</b>	107	116	<b>9</b>	<b>74</b>	<b>77</b>	<b>6</b>
<b>TLC %</b>	95	97	<b>2</b>	73	72	<b>-1</b>	72	72	<b>0</b>	83	73	<b>-10</b>	98	109	<b>11</b>	<b>80</b>	<b>82</b>	<b>2</b>
<b>DLCO%</b>	54	51	<b>-3</b>	NA	54	<b>NA</b>	60	45	<b>-15</b>	55	56	<b>1</b>	70	67	<b>-3</b>	<b>61</b>	<b>65</b>	<b>-3</b>

PT= patient, BL= baseline, Wk16=week 16, Δ= change, FVC= forced vital capacity, FEV1= forced expiratory volume in 1 second, TLC=total lung capacity, DLCO= diffusion capacity, %=percent, FVC, FEV1, and TLC were measured in liters, % of predicted is reported. DLCO was measured in milliliters/minute/millimeter of mercury and reported in % predicted. NA- technically inadequate/not available, \*\* Patient #9 had a severe respiratory infection at the time of this test. All other testing (6MWD, ETT, Echo, Labs, RHC) were delayed 4 weeks until resolution of this infection.