**TABLE 1. DEMOGRAPHIC CHARACTERISTICS AND PULMONARY FUNCTION OF THE PILOT STUDY**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Characteristics** | **Patient 1**  **Pre Post** | | **Patient 2**  **Pre Post** | | **Patient 3**  **Pre Post** | |
| Age, years | 28 | 30 | 34 | 36 | 58 | 60 |
| BMI, kg/m2 | 23.1 | 25.7 | 22.5 | 23.1 | 23.7 | 24.1 |
| Pseudomonas aeruginosa infection, % | yes | yes | yes | yes | yes | yes |
| Staphylococcus aureus infection, % | yes | yes | no | no | no | no |
| Burkholderia cepacia infection | no | no | no | no | yes | yes |
| Pancreatic insufficiency | yes | yes | yes | yes | yes | yes |
| Diabete CF-related | no | no | no | no | yes | yes |
| N of pulmonary exacerbation | 2 | 0 | 4 | 2 | 3 | 2 |
| Lung function | | | | |  |  |
| FEV1, L | 2.55 | 2.11 | 1.58 | 1.16 | 2.08 | 1.58 |
| FEV1, % of predicted | 65.3 | 57 | 43.4 | 34 | 56.1 | 45 |
| FVC, L | 4.52 | 4.23 | 2.7 | 2.18 | 3.9 | 3.28 |
| FVC, % of predicted | 99.6 | 97 | 63.6 | 55 | 83.9 | 73 |
| FEV1/FVC, % | 56.4 | 50 | 58.5 | 53.4 | 53.2 | 48 |
| FEF 25-75%, L/sec | 1.14 | 0.82 | 0.71 | 0.45 | 0.89 | 0.70 |
| FEF 25-75%, % of predicted | 23.4 | 18 | 15.4 | 11 | 22.5 | 19 |
| TLC, L | 7.06 | 7.15 | 4.66 | 5.69 | 7.27 | 7.55 |
| TLC, % of predicted | 115.7 | 117 | 79.4 | 98 | 99.6 | 103 |
| RV, L | 2.21 | 2.53 | 1.85 | 2.79 | 2.3 | 3.42 |
| RV, % of predicted | 151.6 | 159 | 120 | 167 | 100 | 140 |
| RV/TLC, % | 31.3 | 35.4 | 39.7 | 49 | 31.6 | 45.4 |
| RV/TLC, % of predicted | 134.3 | 138 | 157.3 | 175 | 91.3 | 121 |
| FRC, L | 3.79 | 3.24 | 2.17 | 3.12 | 2.6 | 4.28 |
| FRC, % of predicted | 126.9 | 107 | 73.3 | 104 | 72.2 | 117 |
| DLCO, ml/min/mmHg | 22.14 | 30.07 | 27.47 | 23.59 | 26.16 | 20.73 |
| DLCO, % of predicted | 69.1 | 97 | 91.4 | 83 | 83.6 | 69 |

*Definition of abbreviations*: BMI = body mass index; FEV1 = forced expiratory volume in one second; FVC = forced vital capacity; FEV1/FVC = forced expiratory volume in one second and forced vital capacity ratio; TLC = total lung capacity;

**TABLE 2. DAILY PHYSICAL ACTIVITY OF THE PILOT STUDY**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Variable** | **Patient 1**  **Pre Post** | | **Patient 2**  **Pre Post** | | **Patient 3**  **Pre Post** | |
| Total energy expenditure, kcal | 3093 | 2837 | 2509 | 2751 | 2542 | 2996 |
| Active Energy expenditure, kcal | 1377 | 1178 | 752 | 1165 | 420 | 1018 |
| Duration Physical Activity, min/day | 289 | 280 | 197 | 314 | 89 | 192 |
| Average METs, kcal·kg−1·h−1 | 2.1 | 1.9 | 1.8 | 1.9 | 1.4 | 1.7 |
| Steps, number/day | 14164 | 8949 | 8904 | 7711 | 6542 | 5770 |
| Mild intensity activities, min/day | 213 | 241 | 167 | 308 | 84 | 159 |
| Moderate intensity activities, min/day | 60 | 38 | 29 | 6 | 4 | 31 |
| Vigorous intensity activities, min/day | 16 | 1 | 1 | 0 | 1 | 2 |
| Moderate+Vigorous intensity activities, min/day | 76 | 39 | 30 | 6 | 5 | 33 |

*Definition of abbreviations*: METs = Metabolic Equivalents of Task.

**TABLE 3.** **MEASUREMENTS AT SYMPTOM-LIMITED INCREMENTAL CYCLE EXERCISE**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Characteristics** | **Patient 1**  **Pre Post** | | **Patient 2**  **Pre Post** | | **Patient 3**  **Pre Post** | |
| Exercise time (sec) | 540 | 510 | 360 | 345 | 420 | 405 |
| Power *max* (watt) | 190 | 190 | 130 | 130 | 160 | 105 |
| VꞌO2 *peak* (ml/min) | 1890 | 2440 | 1439.2 | 1847 | 1566 | 1653 |
| VꞌO2 *peak*/Kg (ml/min/Kg) | 30 | 34.9 | 24.39 | 30.8 | 20.34 | 21.2 |
| VꞌO2 *peak* (% pred.) | 66.3 | 88 | 55 | 78 | 61 | 73 |
| VꞌO2 @ AT (ml/min) | 1170.4 | 1347 | 1181.1 | 1254 | 962.6 | 1189 |
| VꞌO2 @ AT (ml/min/Kg) | 18.6 | 19.2 | 20 | 20.9 | 12.5 | 15.2 |
| VꞌO2 @ AT (% pred.) | 40.8 | 48.6 | 45 | 52.9 | 37.4 | 52.5 |
| Heart Rate *max* (bpm) | 156 | 161 | 172 | 175 | 160 | 131 |
| Heart Rate *max* (% pred.) | 79.6 | 85 | 90 | 95 | 95 | 82 |
| VꞌO2/HR (ml/beat) | 12.1 | 15.2 | 8.36 | 10.6 | 9.8 | 12.6 |
| Respiratory Rate *max* (bpm) | 49.9 | 39.7 | 58.6 | 57.8 | 42.5 | 32.7 |
| VE *max* (l/min) | 74.9 | 81.2 | 59.3 | 64.4 | 67 | 49 |
| VE/VꞌCO2 @ AT | 33.9 | 31 | 31.7 | 33.4 | 35 | 32.9 |
| VE/VꞌCO2 *peak* | 31.5 | 30.7 | 34 | 33.2 | 33.6 | 33.2 |
| PETCO2 @ AT (mmHg) | 36 | 42 | 38 | 38 | 32 | 38 |
| PETCO2 *peak* (mmHg) | 41 | 42 | 36 | 36 | 34 | 39 |
| Borg score *peak* | 4 | 4 | 7 | 7 | 5 | 5 |
| Leg score *peak* | 5 | 7 | 7 | 7 | 6 | 6 |
| Borg score @ AT | 0.5 | 0.5 | 0 | 0.5 | 3 | 3 |
| Leg score @ AT | 0.5 | 3 | 0.5 | 0.5 | 3 | 4 |
| Δ IC (l) | -0.33 | -0.18 | -0.4 | n.d. | -0.91 | +0.11 |
| VT *peak* (l) | 1.5 | 2 | 1 | 1.1 | 1.58 | 1.5 |
| BR (%) | 21.1 | 3.8 | 0 | -38 | 19.8 | 22.4 |
| ΔSpO2 *peak-rest* | -3 | -4 | -2 | -7 | -5 | -3 |

*Definition of abbreviations*: V’O2 = oxygen uptake ; HR **=** heart rate; SpO2 = arterial oxygen saturation; AT = anaerobic threshold; VT= tidal volume; V’E = minute ventilation; BR = breathing reserve; V’E/V’CO2 = ventilatory equivalent for carbon dioxide; PETCO2= partial pressure of end-tidal CO2; IC = inspiratory capacity; VT= tidal volume; BR = breathing reserve TLC = total lung capacity.

Data recorded at peak exercise.