Supplementary Material

|  |  |  |  |  |  |  |  |  |  |  |  |  |
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|  | **N** | **Pre-T0** | **N** | **T0** | **T1** | **T0 vs T1** | **N** | **T2** | **Pre-T0 vs T2** | **N** | **T3** | **T0 vs T3** |
| HOSPITALIZATIONS | 16 | 0.75±0.58 |  |  |  |  |  |  |  | 16 | 0.31±0.48 | **0.02** |
| PULMONARY EXACERBATIONS | 16 | 1.44±1.59 |  |  |  |  |  |  |  | 16 | 0.50±0.89 | 0.08 |
| SIX MINUTE WALKING TEST | 11 | 560.67±80.87 |  |  |  |  |  |  |  | 11 | 542.95±96.99 | 0.96 |
| CHLORIDE SWEAT TEST | 6 | 114.83±13.73 |  |  |  |  |  |  |  | 6 | 52.00±22.64 | **0.03** |
| BODY MASS INDEX |  |  | 15 | 20.91±2.47 | 21.49±2.43 | **0.01** | 14 | 21.44±2.46 | **0.01** | 14 | 21.20±2.90 | 0.21 |
| FEV1 |  |  | 16 | 2326.9±871.8 | 2696.9±944.15 | **0.0001** | 15 | 2720.7±953.11 | **0.001** | 14 | 2803.57±937.4 | **0.001** |
| FVC |  |  | 16 | 3110.0±936.2 | 3463.1±919.2 | **0.004** | 15 | 3524.0±920.3 | **0.006** | 14 | 3586.3±936.4 | **0.004** |
| FEV1% |  |  | 16 | 72.00±24.27 | 83.19±24.46 | **0.001** | 15 | 84.47±24.41 | **0.001** | 14 | 87.21±21.42 | **0.001** |
| TOTAL DAILY INSULIN DOSE |  |  | 14 | 0.29±0.21 | 0.20±0.20 | 0.06 | 13 | 0.19±0.19 | **0.05** | 12 | 0.21±0.21 | 0.21 |
| HBA1C |  |  | 13 | 6.25±0.75 | 6.01±0.59 | **0.005** | 15 | 6.09±0.64 | 0.29 | 12 | 6.09±0.66 | 0.19 |

TABLE S1: Pulmonary function and metabolic changes in patients affected by cystic fibrosis related diabetes with genotype F508del/F508del before and after treatment with elexacaftor/tezacaftor/ivacaftor. Pre-T0: 1 year before; T0: treatment initiation; T1: 3 months after; T2: 6 months after; T3 1 year after. FEV1: forced expiratory volume in the first second; FVC forced vital capacity; HBA1c: glycated hemoglobin.

**TABLE S2: Pulmonary function and metabolic changes in patients affected by cystic fibrosis related diabetes with genotype F508del/any other CFTR mutation before and after treatment with elexacaftor/tezacaftor/ivacaftor.** Pre-T0: 1 year before; T0: treatment initiation; T1: 3 months after; T2: 6 months after; T3 1 year after. FEV1: forced expiratory volume in the first second; FVC forced vital capacity; HBA1c: glycated hemoglobin.

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| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
|  | **N** | **Pre-T0** | **N** | **T0** | **T1** | **T0 vs T1** | **N** | **T2** | **Pre-T0 vs T2** | **N** | **T3** | **T0 vs T3** |
| HOSPITALIZATIONS | 12 | 1.00±1.21 |  |  |  |  |  |  |  | 12 | 0.33±0.49 | 0.07 |
| PULMONARY EXACERBATIONS | 12 | 1.92±1.62 |  |  |  |  |  |  |  | 12 | 0.42±0. 67 | **0.01** |
| SIX MINUTE WALKING TEST | 8 | 592.75±43.35 |  |  |  |  |  |  |  | 8 | 613.71±74.07 | 0.57 |
| CHLORIDE SWEAT TEST | 9 | 121.00±26.99 |  |  |  |  |  |  |  | 9 | 48.56±15.14 | **0.008** |
| BODY MASS INDEX |  |  | 11 | 22.82±3.15 | 23.95±3.48 | **0.004** | 12 | 23.69±3.25 | **0.01** | 11 | 24.10±3.54 | **0.005** |
| FEV1 |  |  | 11 | 2555.5±1276.5 | 2906.4±981.87 | **0.04** | 12 | 2842.5±1076.7 | **0.04** | 11 | 2730.9±1048.4 | 0.18 |
| FVC |  |  | 11 | 3496.4±1190.9 | 3897.3±843.6 | 0.09 | 12 | 3908.3±1005.0 | **0.03** | 11 | 3780.0±975.6 | 0.11 |
| FEV1% |  |  | 11 | 81.18±35.78 | 88.73±23.29 | 0.19 | 12 | 86.50±27.64 | 0.33 | 11 | 83.73±26.07 | 0.72 |
| TOTAL DAILY INSULIN DOSE |  |  | 11 | 0.22±0.23 | 0.15±0.17 | 0.12 | 12 | 0.22±0.22 | 0.40 | 11 | 0.22±0.25 | 0.26 |
| HBA1C |  |  | 10 | 6.07±0.45 | 5.84±0.57 | 0.06 | 11 | 5.62±0.40 | **0.004** | 11 | 5.77±0.51 | **0.02** |