|  |  |  |
| --- | --- | --- |
| Drug | ivacaftor/lumacaftor (IVA/LUM) | ivacaftor/tezacaftor (IVA/TEZ) |
| Month | 0 | 1 | 3 | 0 | 1 | 3 |
| Genotype | Phe508del/ Phe508del | Phe508del/ Phe508del |
| Female (n) | 6 (13) | 4 (8) |
| Age, years | **30** (23-41) | **41** (28-50) |
| ppFEV1 | **29** (12-44) | **37** (12-44) | **33** (17-42) | **26** (19-35) | **26** (19-38) | **29** (19-40) |
| ppFVC  | **43** (31-65) | **48** (31-65) | **44** (30-66) | **43** (36-80) | **47** (35-78) | **48**(29-74) |
| Weight, kg | **55**(48-90) | **55**(48-90) | **55** (47-92) | **58**(48-73) | **59**(50-75) | **58**(47-75) |
| BMI | **20**(16-28) | **20** (18-23) | **20** (18-29) | **22**(21-24) | **21**(20-24) | **22** (21-24) |
| CRP mg/ L | **5** (5-31) | **15** (5-44) | **8.5** (5-149) | **23** (5-77) | **23** (5-45) | **29** (5-51) |
| WBC | **9.3** (6.7-11.6) | **10.7**(6.8-17.1)  | **9.1**(5.1-13.8) | **9.6** (4.8-15.3) | **11.5**(6.1-17.6) | **10.6** (5.2-16.3) |
| Neutrophil | **6.2** (5.2-10.1) | **8.8** (4.9-12.7) | **6.6** (3.2-14.1) | **7.0**(2.9-12.8) | **9.6** (3.5-15.9) | **8.3** (2.6-13.7) |

Supplementary file 1

Supplementary file 1. Demographic and clinical characteristics for CF patients on ivacaftor/lumacaftor (IVA/LUM) and ivacaftor/tezacaftor (IVA/TEZ). Data are expressed as median and range. BMI: Body Mass Index; ppFEV: percent predicted forced expiratory volume, ppFVC: forced vital capacity, CRP: C-reactive protein. WBC: white blood count.