



Figure S1. CFTR-dependent chloride efflux in circulating mononuclear cells. CFTR-dependent chloride efflux is shown in MNCs obtained from each CF patient at baseline (0), 1, 6, and 12 months from the beginning of Orkambi® treatment. While all patients are homozygous for the *F508del* mutation, patients #4 and patient #13 have also the *A238V* mutation in cis.