

## SUPPLEMENTARY TABLES

**Supplementary Table 1. List of active substances administered as inhaled corticosteroids (ICS).**

Active substance(s)	No. (%)
Budesonide (ICS)/Formoterol fumarate dihydrate (LABA)	39 (55%)
Fluticasone propionate (ICS)/Salmeterol (LABA)	22 (31%)
Fluticasone propionate (ICS)	9 (13%)
Beclometasone dipropionate (ICS) /formoterol fumarate dihydrate (LABA)	1 (1%)

In total, 70 out of 168 included patients were prescribed ICS. In most cases, it concerned a drug combining a long-acting bronchodilator (LABA) and an inhaled corticosteroid (ICS).

**Supplementary Table 2. Genotype-specific non-linear age association with LTL attrition in CF patients (n=85) correction for regression to the mean.**

Genotype	Age	$\beta$ (95% CI)	P-value
Homozygous	1	0.022 (0.005, 0.039)	0.011
Heterozygous	1	0.001 (-0.013, 0.014)	0.93
Homozygous	5	0.013 (0.002, 0.024)	0.018
Heterozygous	5	-0.001 (-0.011, 0.009)	0.89
Homozygous	17	-0.012 (-0.022, -0.002)	0.021
Heterozygous	17	-0.004 (-0.009, 0.001)	0.084

Estimates presented with 95%CI representing the association between  $\Delta$ LTL (with correction for regression to the mean as postulated by Verhulst and colleagues [2]) and age at TP1 for each year increment at different ages (1, 5, and 17 years, based on 25<sup>th</sup>, median and 75<sup>th</sup> percentile of the age distribution). Models adjusted for sex and time between samples within an individual. P-interaction between the quadratic term of age at TP1 and genotype ( $P=0.039$ ), reflecting the genotype-specific non-linear LTL attrition-age association. In total 44 patients were homozygous for the  $\Delta$ F508 mutation and 41 were heterozygous.