

**Table S1.** Spearman correlation analysis between bromatological and biochemical data at baseline

		<b>BMI</b> <b>(kg/m<sup>2</sup>)</b>	<b>Vit A</b> <b>(µg/dL)</b>	<b>Album</b> <b>(g/dL)</b>	<b>CHOL</b> <b>(mg/dL)</b>	<b>LDL</b> <b>(mg/dL)</b>	<b>CHOL/</b> <b>HDL ratio</b>	<b>no-HDL CHOL</b> <b>(mg/dL)</b>	<b>LDL/HDL</b> <b>ratio</b>
<b>Kcal/day</b>	r <sub>s</sub>	-0.198	0.151	-0.357	-0.536	-0.364	0.227	-0.382	0.073
	p	0.559	0.699	0.281	0.089	0.272	0.502	0.247	0.832
<b>Protein</b> <b>(g)</b>	r <sub>s</sub>	-0.032	0.088	-0.002	-0.009	-0.005	0.200	-0.009	0.091
	p	0.925	0.821	0.995	0.979	0.989	0.555	0.979	0.790
<b>Lipids</b> <b>(g)</b>	r <sub>s</sub>	-0.284	-0.176	-0.266	-0.551	-0.465	-0.205	-0.538	-0.196
	p	0.397	0.651	0.429	0.079	0.150	0.545	0.088	0.564
<b>Carb (g)</b>	r <sub>s</sub>	-0.268	0.756	-0.163	-0.451	-0.105	0.497	-0.164	0.392
	p	0.426	<b>0.018</b>	0.632	0.164	0.759	0.120	0.630	0.233
<b>Fibers</b> <b>(g)</b>	r <sub>s</sub>	0.196	0.662	-0.095	-0.111	0.005	0.346	0.041	0.230
	p	0.563	0.052	0.781	0.746	0.989	0.298	0.904	0.495
<b>Calcium</b> <b>(mg)</b>	r <sub>s</sub>	0.143	0.268	-0.279	-0.591	-0.491	0.073	-0.491	-0.064
	p	0.675	0.486	0.406	0.056	0.125	0.832	0.125	0.853
<b>Chol</b> <b>(mg)</b>	r <sub>s</sub>	-0.074	-0.201	-0.151	-0.445	-0.555	-0.282	-0.536	-0.318
	p	0.829	0.604	0.658	0.170	0.077	0.401	0.089	0.340
<b>SFA</b> <b>(g)</b>	r <sub>s</sub>	-0.320	-0.272	0.130	-0.221	-0.355	-0.571	-0.396	-0.424
	p	0.337	0.478	0.703	0.513	0.284	0.066	0.228	0.194
<b>MuFA</b> <b>(g)</b>	r <sub>s</sub>	0.092	-0.055	-0.041	-0.350	-0.613	-0.805	-0.628	-0.676
	p	0.787	0.888	0.905	0.292	<b>0.045</b>	<b>0.003</b>	<b>0.039</b>	<b>0.023</b>
<b>PuFA (g)</b>	r <sub>s</sub>	-0.189	0.277	0.420	-0.292	-0.359	-0.541	-0.441	-0.426
	p	0.577	0.471	0.199	0.383	0.278	0.085	0.175	0.191

**Table S2.** Spearman correlation analysis between bromatological and biochemical data at 12 months

		BMI (kg/m <sup>2</sup> )	Vit A (µg/dL)	Album (g/dL)	CHOL (mg/dL)	LDL (mg/dL)	CHOL/ HDL ratio	no-HDL CHOL (mg/dL)	LDL/HDL ratio
Kcal/day	r <sub>s</sub>	-0.437	-0.400	0.571	-0.101	-0.164	-0.591	-0.487	-0.638
	p	0.179	0.286	0.066	0.768	0.631	0.056	0.128	<b>0.035</b>
Protein (g)	r <sub>s</sub>	-0.513	-0.251	0.664	0.354	0.096	-0.224	-0.146	-0.137
	p	0.107	0.515	<b>0.026</b>	0.285	0.779	0.508	0.667	0.687
Lipids (g)	r <sub>s</sub>	-0.308	-0.583	0.350	-0.128	-0.182	-0.373	-0.415	-0.487
	p	0.357	0.099	0.291	0.707	0.593	0.259	0.205	0.128
Carb (g)	r <sub>s</sub>	-0.363	-0.083	0.470	-0.252	-0.382	-0.773	-0.642	-0.797
	p	0.272	0.831	0.145	0.455	0.247	<b>0.005</b>	<b>0.033</b>	<b>0.003</b>
Fibers (g)	r <sub>s</sub>	-0.023	-0.100	-0.076	-0.218	-0.173	0.387	0.009	0.409
	p	0.946	0.798	0.824	0.520	0.611	0.239	0.979	0.212
Calcium (mg)	r <sub>s</sub>	-0.525	-0.234	-0.152	-0.216	-0.282	0.068	-0.137	0.023
	p	0.097	0.544	0.655	0.524	0.400	0.842	0.688	0.947
Chol (mg)	r <sub>s</sub>	-0.830	-0.519	0.453	0.119	-0.146	-0.401	-0.326	-0.438
	p	<b>0.002</b>	0.152	0.162	0.727	0.669	0.222	0.327	0.177
SFA (g)	r <sub>s</sub>	-0.567	-0.667	0.292	0.005	-0.014	-0.197	-0.144	-0.229
	p	0.069	0.050	0.383	0.989	0.968	0.562	0.672	0.498
MuFA (g)	r <sub>s</sub>	-0.125	-0.815	0.193	0.118	0.394	0.430	0.369	0.431
	p	0.714	0.007	0.571	0.731	0.231	0.187	0.264	0.185
PuFA (g)	r <sub>s</sub>	0.131	-0.009	0.257	-0.014	0.157	-0.147	0.122	-0.049
	p	0.702	0.983	0.446	0.968	0.645	0.665	0.720	0.887

**Table S3.** Protein and metabolic changes after one year of ETI treatment in patients with CF carrying Minimal Function (MF) heterozygosity with Phe508del mutation

Genotype group	Class of defect	Protein changes	Albumin changes (%)		non-HDL CHOL changes (%)	
			average (SD)	min-max	average (SD)	min-max
First group (n = 5)	I	early stop truncation	15 (10)	0 – 24	18 (51)	-28 – 105
Second group (n = 9)	I	late stop truncation and other MF defects	19 (5)	5 – 29	59 (40) *	4 – 129
Third group (n = 6)	II	Other not responsive MF defects <sup>a</sup>	11 (10)	0 – 29	69 (46) *	8 – 159

\*p < 0.05 vs first group. <sup>a</sup> Not responsive to ivacaftor or tezacaftor. ETI: elexacaftor/tezacaftor/ivacaftor