

Table 1: median baseline values and quartiles per medication group

	Lumacaftor/ivacaftor (N=23)	Ivacaftor (N=7)
Age (Q1-Q3)	19 (15-34)	14 (9-18)
Sex (number male/ number female)	21 male, 22 female	3 male, 4 female
FEV₁ percentage predicted (Q1-Q3)	62 (35-81)	77 (65-113)
BMI (Q1-Q3)	19.8 (15.3-23.3)	19.0 (14.6-25.4)
Exacerbations in the year before start (Q1-Q3)	2 (0.75-5)	0 (0-3)
Brody score, total without airtrapping; score out of 100 (Q1-Q3)	20.60 (15.97-29.75)	9.95 (1.39-22.11)
Brody subscore: bronchiectasis; score out of 100 (Q1-Q3)	31.25 (17.36-41.67)	17.36 (2.78-34.03)
Brody subscore: mucus plugging; score out of 100 (Q1-Q3)	19.44 (11.11-27.78)	8.33 (0-19.44)
Brody subscore: peribronchial thickening; score out of 100 (Q1-Q3)	24.07 (19.44-31.02)	11.11 (1.85-14.07)
Brody subscore: parenchyma; score out of 100 (Q1-Q3)	9.26 (1.85-12.96)	1.85 (0-5.56)