

Table 1. Demographic and clinical characteristics of the patients.

Results are shown as mean \pm SD and ranges or numbers of patients and percentile.

		Range (Median)
N	89	
Females, n (%)	42 (47)	
Age, years	17.6 \pm 9.0	4.25-49.0 (16.0)
BMI %tile	57.2 \pm 27.45	4.1-98.9 (55.2)
CFTR Genotype:		
2 mutations from class I-III, n (%)	62 (70)	
1 or 2 mutations from class IV-V, n (%)	25 (27)	
1 mutation unknown, n (%)	2 (3)	
<i>P. aeruginosa</i> chronic colonization, n (%)	19 (21)	
Pancreatic insufficiency, n (%)	61 (69)	
CF related diabetes, n (%)	11 (12)	
Number of PEx previous year		
Minor	1.0 \pm 1.0	0-5 (1.0)
Major	0.3 \pm 0.6	0-2 (0)
Abx treatment days previous year		
PO	25.8 \pm 29.3	0-168 (14)
IV	7.1 \pm 15.3	0-112 (0)
FEV ₁ , % predicted	97 \pm 12	80-138 (96)
FEV ₁ z-score	-0.26 \pm 1.0	-1.7-2.7 (-0.26)
LCI	10.15 \pm 2.5	6.0-18.9 (10.4)
Chest HRCT Brody score (n=65)		
Bronchiectasis sub-score	4.9 \pm 8.4	0-33.25 (1.0)
Mucous plugging sub-score	4.1 \pm 4.4	0-18 (2.0)
Peribronchial thickening sub-score	7.6 \pm 7.7	0-32 (6.0)
Parenchyma sub-score	0.8 \pm 1.5	0-8 (0)
Air trapping sub-score	5.25 \pm 4.7	0-18 (5.0)
Total Brody score	22.9 \pm 21.0	0-104.5 (17.5)
6MWD, meters (n=68)	562 \pm 95	360-787 (580)
6MWD z-score (n=68)	-1.9 \pm 1.4	-5.0-1.0 (-1.8)

Abbreviations: BMI: body mass index; CFTR: Cystic fibrosis transmembrane conductance regulator; PEx: pulmonary exacerbation; Abx: antibiotic; PO: per os; IV: intravenous; FEV₁: forced expiratory volume in 1 second; LCI: lung clearance index; HRCT: high-resolution computed tomography; 6MWD: 6-minute walk distance.

Table 2. Associations between FEV₁ z-score, LCI, HRCT-TBS and CF severity markers.

		n (%)	FEV ₁ z-score (mean±SD)	P- value	LCI (mean±SD)	P-value	HRCT- TBS (mean± SD)	P-value
CFTR mutation classes	<i>2 mutati ons from class I-III</i>	62 (71)	-0.4 ± 1.0	0.36	10.4 ± 2.9	0.87	23.9 ± 19.5	0.59
	<i>1 or 2 mutati ons from class IV-V</i>	25 (29)	-0.17 ± 0.9		10.3 ± 2.5		20.8 ± 24.5	
Pancreatic status	<i>PI</i>	61 (69)	-0.35 ± 1.1	0.65	10.5 ± 2.9	0.52	23.6 ± 19.4	0.69
	<i>PS</i>	28 (31)	-0.24 ± 1.0		10.0 ± 2.4		21.4 ± 25.1	
PA colonization	<i>Pos.</i>	19 (21)	1.0 ± 0.8	0.08	11.9 ± 3.1	0.01	38.3±25. 0	0.01
	<i>Neg.</i>	70 (79)	-0.17 ± 1.0		9.7 ± 2.3		20.2 ± 19.4	
CFRD	<i>Pos.</i>	11 (12)	-0.6 ± 1.2	0.36	11.1 ± 3.8	0.36	11.4± 12.5	0.16
	<i>Neg.</i>	78 (88)	-0.28 ± 1.0		10.2 ± 2.5		24.1± 21.5	
No. of minor	0	41 (46)	0.04± 0.94		9.8 ± 3.2		21.6±	

PEx				0.22		0.18	20.6	0.61
	1≤	48 (54)	-0.43 ± 1.0		10.4 ± 2.3		24.3± 21.7	
PO Abx treatment days	≤14	42 (47)	0.04± 0.94	0.22	9.8 ± 3.2	0.18	21.6± 20.6	0.61
	14<	47 (53)	-0.43 ± 1.0		10.4 ± 2.3		24.3 ± 21.7	
No. of major PEx	0	61 (69)	0.08± 0.98	0.0002	10.0± 2.75	0.04	18.1± 15.3	0.0007
	1≤	28 (31)	-0.75± 0.91		11.6 ± 2.9		37.8± 28.7	
IV Abx treatment days	≤14	76 (85)	-0.1 ± 0.98	0.0006	9.9 ± 2.4	0.03	20.2± 19.5	0.002
	14<	13 (15)	-0.75± 0.93		11.2 ± 2.6		45.4± 20.9	

Abbreviations: FEV₁: forced expiratory volume in one second; LCI: lung clearance index; PI: pancreatic insufficiency; HRCT: high resolution computed tomography; TBS: total Brody score; PS: pancreatic sufficiency; PA: *Pseudomonas aeruginosa*; CFRD: CF-related diabetes; No.: number; PEx: pulmonary exacerbations; PO: per os; Abx: antibiotic; IV: intravenous.