

Table 1

Parameter	All patients	Nintedanib	Pirfenidone	No treatment	P-values
N (%)	178	62 (34.8%)	80 (44.9%)	36 (20.2%)	
Age, mean (SD)	73.80 (7.53)	72.16 (7.15)	73.66 (6.87)	76.92 (8.74)	0.0097
Men, n (%)	136 (76.4%)	47 (75.8%)	62 (77.5%)	27 (75.0%)	0.95
BMI, mean (SD)	27.31 (4.51)	27.62 (4.62)	27.57 (4.16)	26.16 (5.02)	0.24
FVC (L), mean (SD)	3.04 (0.86)	3.05 (0.91)	3.08 (0.83)	2.94 (0.88)	0.7
FVC (% pred), mean (SD)	89.51 (19.52)	88.48 (19.57)	90.09 (18.71)	90.00 (21.65)	0.878
DLCO (% pred), mean (SD)	52.85 (13.25)	53.81 (12.68)	52.63 (13.13)	51.58 (14.80)	0.725
Change in FVC % pred. from baseline to 12 months, mean (SD)	0.13 (10.16)	0.20 (9.92)	0.06 (10.44)	0.15 (10.41)	0.997
Change in DLCO % pred. from baseline to 12 months, mean (SD)	-5.03 (7.98)	-4.70 (6.83)	-5.37 (9.10)	-4.85 (7.34)	0.9
6MWT (meters), mean (SD)	442.61 (105.58)	475.92 (95.20)	436.09 (106.85)	390.97 (101.61)	0.0009
Smoking status, n (%)					0.039
Never	46 (25.8%)	10 (16.1%)	25 (31.2%)	11 (30.6%)	
Active	11 (6.2%)	1 (1.6%)	6 (7.5%)	4 (11.1%)	
Former	121 (68.0%)	51 (82.3%)	49 (61.2%)	21 (58.3%)	
GAP index, n (%)					0.08
I	88 (49.7%)	34 (54.8%)	36 (45.0%)	18 (51.4%)	
II	82 (46.3%)	27 (43.5%)	42 (52.5%)	13 (37.1%)	
III	7 (4.0%)	1 (1.6%)	2 (2.5%)	4 (11.4%)	
Progression at 12 months, n (%)	84 (47.2%)	27 (43.5%)	38 (47.5%)	19 (52.8%)	0.67

Table 1: Baseline characteristics

SD standard deviation, BMI Body Mass Index, FVC forced vital capacity, DLCO diffusion capacity for carbon monoxide, 6MWT six-minute walk test, GAP index Gender-Age-Physiology index, Disease progression was defined as an absolute decline in the percentage of predicted FVC \geq 5% points and/or an absolute decline in the percentage of predicted DLCO \geq 10% points and/or all-cause mortality within 12 months.

Fig. 1

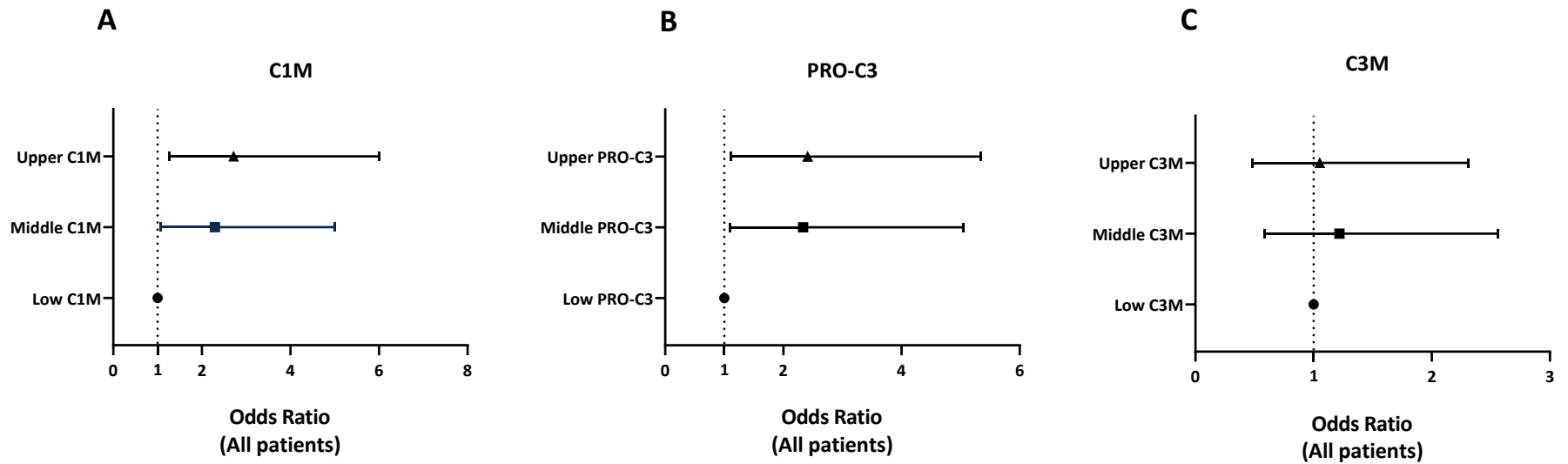


Figure 1: Risk of disease progression at 12 months for IPF patients.

Odds ratio from IPF patients divided into tertiles from baseline biomarker of C1M (A), PRO-C3 (B) and C3M (C) are shown for the middle and upper tertile compared to the lowest tertile. Disease progression was defined as $\geq 5\%$ decline in FVC and/or $\geq 10\%$ decline in DLco or all-cause mortality at 12 months. Data are presented as mean and 95% CI (error bars) adjusted for age, sex, and baseline levels of FVC and DLco. Each tertile had $n=59-60$.

Fig. 2

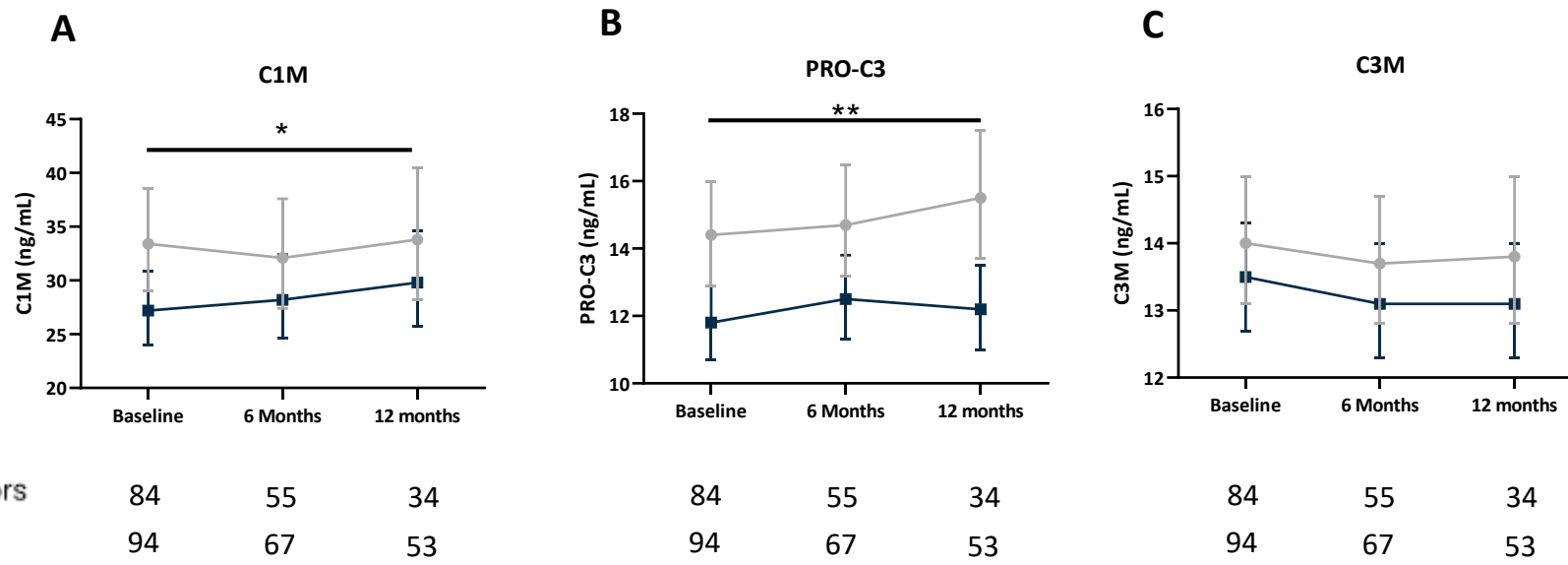


Figure 2: Longitudinal biomarker levels are elevated in progressive IPF patients.

Serum levels of C1M (A), PRO-C3 (B) and C3M (C) are shown at baseline, six months and 12 months for stable (dark blue) and progressive (grey) patients with IPF. Disease progression was defined as $\geq 5\%$ decline in FVC and/or $\geq 10\%$ decline in DLco or all-cause mortality within 12 months. Data are presented as mean and 95% CI (error bars) adjusted for age and sex. The number of evaluable samples available for analysis at each time point is provided in the graph. The P-values for the interaction between visit and progression status for C1M ($P=0.63$), PRO-C3 ($P=0.46$) and for C3M ($P=0.86$). Significant differences between progressive and stable patients over one year are shown as ** ($P<0.01$) and * ($P<0.05$).

Fig. 3

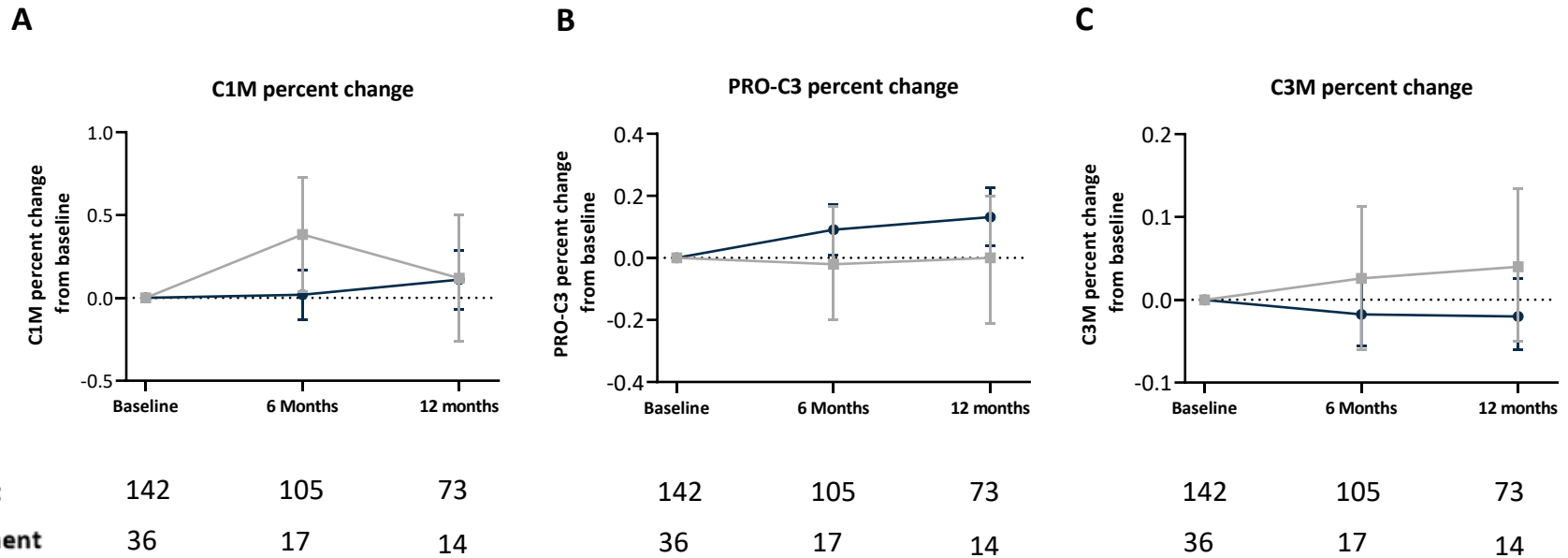


Figure 3: Change from baseline over time in type I and III collagen turnover in treated and untreated IPF patients. Percent change from baseline in C1M (A), PRO-C3 (B) and, C3M (C) at six months and 12 months for treated (nintedanib/pirfenidone) (Dark blue) and non-treated (grey) patients with IPF. Data are presented as mean and 95% CI (error bars) adjusted for age, sex and baseline levels of C1M, PRO-C3 or C3M. The number of evaluable samples available for analysis at each time point is provided in the graph. The P-values for the interaction between visit and treatment status for C1M ($P=0.18$), PRO-C3 ($P=0.82$) and for C3M ($P=0.79$).