

Supplementary Table I. Correlation of the 6MWD at baseline with the clinical parameters at baseline.

	N	correlation coefficient r or q^* (p)
Length V0	165	0.227 (p=0.003)
Weight V0	165	-0.095 (p=0.227)
Dis dur Rayn V0	162	-0.086 (p=0.275)
Dis dur nonRayn V0	137	0.007 (p=0.938)
Age V0	165	-0.404 (p<0.001)
mRSS V0	165	-0.127 (p=0.104)
ESR V0	164	-0.100 (p=0.202)
CRP V0	165	-0.071 (p=0.367)
HgB V0	165	0.221 (p=0.004)
Creat V0	165	-0.229 (p=0.003)*
CK V0	160	-0.059 (p=0.462)
TLC V0	164	0.129 (p=0.101)
FVC V0	165	0.027 (p=0.733)
FEV1 V0	165	0.120 (p=0.125)
DLCO V0	165	0.081 (p=0.301)
Peak TRV V0	131	-0.177 (p=0.043)

Supplementary Table IIA. Correlation of the 6MWD at 18-month visit with the clinical parameters at baseline and at 18-month visit.**Supplementary Table IIB.** Correlation of the 6MWD at 18-month visit with the clinical parameters at baseline and at 18-month visit in those patients without ILD and without PAH at 18-month visit (after exclusion of those with ILD and PAH at 18-month visit).

Suppl. Table IIA	N	correlation coefficient r or q^* (p)		N	correlation coefficient r or q^* (p)
Length V0	130	0.305 (p<0.001)	Weight V0	130	0.036 (p=0.682)
Dis dur Rayn V0	128	-0.086 (p=0.336)	Dis dur nonRayn V0	112	-0.072 (p=0.448)
6MWD V0	130	0.817 (p<0.001)	Age V0	130	-0.411 (p<0.001)
mRSS V0	130	-0.084 (p=0.341)	mRSS V18	128	-0.013 (p=0.887)
ESR V0	129	-0.116 (p=0.191)	ERS V18	126	-0.182 (p=0.041)
CRP V0	130	-0.109 (p=0.219)	CRP V18	130	-0.294 (p=0.001)
HgB V0	130	0.331 (p<0.001)	HgB V18	130	0.329 (p<0.001)
Creat V0	130	-0.022 (p=0.802)*	Creat V18	130	-0.024 (p=0.784)*
CK V0	126	-0.026 (p=0.776)	CK V18	129	0.106 (p=0.233)
TLC V0	129	0.176 (p=0.046)	TLC V18	130	0.179 (p=0.041)
FVC V0	130	0.089 (p=0.313)	FVC V18	130	0.051 (p=0.561)
FEV1 V0	130	0.148 (p=0.094)	FEV1 V18	130	0.169 (p=0.055)
DLCO V0	130	0.161 (p=0.067)	DLCO V18	130	0.156 (p=0.076)
Peak TRV V0	106	-0.165 (p=0.092)	Peak TRV V18	109	-0.312 (p=0.001)

Suppl. Table IIB	N	correlation coefficient r or q^* (p)		N	correlation coefficient r or q^* (p)
Length V0	126	0.298 (p=0.001)	Weight V0	126	0.043 (p=0.636)
Dis dur Rayn V0	124	-0.094 (p=0.301)	Dis dur nonRayn V0	108	-0.071 (p=0.463)
6MWD V0	126	0.819 (p<0.001)	Age V0	126	-0.415 (p<0.001)
mRSS V0	126	-0.072 (p=0.426)	mRSS V18	125	0.013 (p=0.885)
ESR V0	125	-0.101 (p=0.264)	ERS V18	122	-0.170 (p=0.061)
CRP V0	126	-0.107 (p=0.235)	CRP V18	126	-0.287 (p=0.001)
HgB V0	126	0.322 (p<0.001)	HgB V18	126	0.322 (p<0.001)
Creat V0	126	-0.046 (p=0.610)	Creat V18	126	-0.034 (p=0.708)
CK V0	122	-0.077 (p=0.400)	CK V18	125	0.105 (p=0.246)
TLC V0	125	0.169 (p=0.060)	TLC V18	126	0.171 (p=0.055)
FVC V0	126	0.091 (p=0.310)	FVC V18	126	0.053 (p=0.558)
FEV1 V0	126	0.158 (p=0.077)	FEV1 V18	126	0.171 (p=0.055)
DLCO V0	126	0.168 (p=0.061)	DLCO V18	126	0.158 (p=0.077)
Peak TRV V0	102	-0.169 (p=0.089)	Peak TRV V18	105	-0.341 (p<0.001)

Supplementary Table IIIA. Correlation of the 6MWD at 30-month visit with the clinical parameters at baseline and at 30-month visit.

Supplementary Table IIIB. Correlation of the 6MWD at 30-month visit with the clinical parameters at baseline and at 30-month visit in those patients without ILD and without PAH at 30-month visit (after exclusion of those with ILD and PAH at 30-month visit).

Suppl. Table IIIA	N	correlation coefficient r or χ^2 (p)		N	correlation coefficient r or χ^2 (p)
Length V0	98	0.209 (p=0.038)	Weight V0	98	0.023 (p=0.824)
Dis dur Rayn V0	97	-0.138 (p=0.178)	Dis dur nonRayn V0	85	-0.017 (p=0.877)
6MWD V0	98	0.745 (p<0.001)	Age V0	98	-0.389 (p<0.001)
mRSS V0	98	-0.077 (p=0.454)	mRSS V30	97	-0.119 (p=0.246)
ESR V0	97	-0.058 (p=0.575)	ERS V30	96	-0.260 (p=0.011)
CRP V0	98	-0.123 (p=0.228)	CRP V30	97	-0.248 (p=0.014)
HgB V0	98	0.223 (p=0.027)	HgB V30	98	0.224 (p=0.027)
Creat V0	98	-0.054 (p=0.601)*	Creat V30	97	-0.150 (p=0.141)*
CK V0	94	0.022 (p=0.836)	CK V30	94	0.186 (p=0.073)
TLC V0	97	0.136 (p=0.184)	TLC V30	98	0.045 (p=0.657)
FVC V0	98	0.005 (p=0.962)	FVC V30	98	0.081 (p=0.426)
FEV1 V0	98	0.065 (p=0.524)	FEV1 V30	98	0.119 (p=0.242)
DLCO V0	98	0.177 (p=0.081)	DLCO V30	98	0.183 (p=0.071)
Peak TRV V0	84	-0.209 (p=0.057)	Peak TRV V30	80	-0.421 (p<0.001)
Suppl. Table IIIB	N	correlation coefficient r or χ^2 (p)		N	correlation coefficient r or χ^2 (p)
Length V0	93	0.195 (p=0.061)	Weight V0	93	0.032 (p=0.762)
Dis dur Rayn V0	92	-0.133 (p=0.205)	Dis dur nonRayn V0	80	-0.018 (p=0.871)
6MWD V0	93	0.744 (p<0.001)	Age V0	93	-0.391 (p<0.001)
mRSS V0	93	-0.053 (p=0.617)	mRSS V30	92	-0.094 (p=0.373)
ESR V0	92	-0.034 (p=0.749)	ERS V30	91	-0.259 (p=0.013)
CRP V0	93	-0.118 (p=0.262)	CRP V30	92	-0.257 (p=0.013)
HgB V0	93	0.212 (p=0.041)	HgB V30	93	0.222 (p=0.033)
Creat V0	93	-0.080 (p=0.443)*	Creat V30	93	-0.165 (p=0.116)*
CK V0	89	-0.048 (p=0.657)	CK V30	90	0.181 (p=0.088)
TLC V0	92	0.118 (p=0.262)	TLC V30	93	0.038 (p=0.721)
FVC V0	93	0.008 (p=0.942)	FVC V30	93	0.078 (p=0.458)
FEV1 V0	93	0.078 (p=0.455)	FEV1 V30	93	0.120 (p=0.250)
DLCO V0	93	0.187 (p=0.073)	DLCO V30	93	0.175 (p=0.094)
Peak TRV V0	79	-0.202 (p=0.075)	Peak TRV V30	75	-0.411 (p<0.001)

Supplementary Table IVA. Correlation of the 6MWD at 42-month visit with the clinical parameters at baseline and at 42-month visit.

Supplementary Table IVB. Correlation of the 6MWD at 42-month visit with the clinical parameters at baseline and at 42-month visit in those patients without ILD and without PAH at 42-month visit (after exclusion of those with ILD and PAH at 42-month visit).

Suppl. Table IVA	N	correlation coefficient r or ρ^* (p)		N	correlation coefficient r or ρ^* (p)
Length V0	77	0.335 (p=0.003)	Weight V0	77	0.036 (p=0.756)
Dis dur Rayn V0	76	-0.276 (p=0.016)	Dis dur nonRayn V0	64	-0.241 (p=0.055)
6MWD V0	77	0.694 (p<0.001)	Age V0	77	-0.486 (p<0.001)
mRSS V0	77	-0.118 (p=0.308)	mRSS V42	77	0.034 (p=0.771)
ESR V0	77	-0.129 (p=0.262)	ERS V42	76	-0.196 (p=0.090)
CRP V0	77	-0.049 (p=0.671)	CRP V42	77	-0.169 (p=0.143)
HgB V0	77	0.343 (p=0.001)	HgB V42	77	0.241 (p=0.035)
Creat V0	77	-0.050 (p=0.669)*	Creat V42	77	-0.037 (p=0.747)*
CK V0	73	-0.008 (p=0.945)	CK V42	77	-0.032 (p=0.784)
TLC V0	76	0.110 (p=0.345)	TLC V42	76	0.126 (p=0.277)
FVC V0	77	-0.098 (p=0.395)	FVC V42	76	-0.030 (p=0.799)
FEV1 V0	77	-0.062 (p=0.590)	FEV1 V42	76	0.026 (p=0.824)
DLCO V0	77	0.036 (p=0.759)	DLCO V42	76	0.067 (p=0.562)
Peak TRV V0	68	-0.136 (p=0.268)	Peak TRV V42	65	-0.501 (p<0.001)
Suppl. Table IVB	N	correlation coefficient r or ρ^* (p)		N	correlation coefficient r or ρ^* (p)
Length V0	74	0.402 (p<0.001)	Weight V0	74	0.049 (p=0.677)
Dis dur Rayn V0	73	-0.269 (p=0.021)	Dis dur nonRayn V0	61	-0.309 (p=0.015)
6MWD V0	74	0.648 (p<0.001)	Age V0	74	-0.443 (p<0.001)
mRSS V0	74	-0.030 (p=0.800)	mRSS V42	74	0.075 (p=0.525)
ESR V0	74	-0.093 (p=0.432)	ERS V42	74	-0.164 (p=0.164)
CRP V0	74	0.003 (p=0.981)	CRP V42	74	-0.071 (p=0.546)
HgB V0	74	0.369 (p=0.001)	HgB V42	74	0.273 (p=0.019)
Creat V0	74	-0.003 (p=0.981)	Creat V42	74	0.017 (p=0.887)
CK V0	71	-0.031 (p=0.796)	CK V42	74	-0.011 (p=0.926)
TLC V0	73	0.098 (p=0.410)	TLC V42	73	0.129 (p=0.277)
FVC V0	74	-0.082 (p=0.487)	FVC V42	73	-0.026 (p=0.826)
FEV1 V0	74	-0.035 (p=0.765)	FEV1 V42	73	0.030 (p=0.799)
DLCO V0	74	0.036 (p=0.761)	DLCO V42	73	0.089 (p=0.454)
Peak TRV V0	65	-0.073 (p=0.566)	Peak TRV V42	62	-0.464 (p<0.001)

Supplementary Table VA. Correlation of the 6MWD at 54-month visit with the clinical parameters at baseline and at 54-month visit.

Supplementary Table VB. Correlation of the 6MWD at 54-month visit with the clinical parameters at baseline and at 54-month visit in those patients without ILD and without PAH at 54-month visit (after exclusion of those with ILD and PAH at 54-month visit).

Suppl. Table VA	N	correlation coefficient r or ρ^* (p)		N	correlation coefficient r or ρ^* (p)
Length V0	65	0.183 (p=0.145)	Weight V0	65	-0.048 (p=0.707)
Dis dur Rayn V0	64	-0.128 (p=0.314)	Dis dur nonRayn V0	52	-0.156 (p=0.270)
6MWD V0	65	0.765 (p<0.001)	Age V0	65	-0.405 (p=0.001)
mRSS V0	65	-0.215 (p=0.085)	mRSS V54	62	-0.063 (p=0.625)
ESR V0	65	-0.020 (p=0.875)	ERS V54	64	-0.123 (p=0.333)
CRP V0	65	-0.042 (p=0.737)	CRP V54	65	-0.155 (p=0.216)
HgB V0	65	0.335 (p=0.006)	HgB V54	65	-0.277 (p=0.026)
Creat V0	65	-0.092 (p=0.467)*	Creat V54	62	0.144 (p=0.265)*
CK V0	63	-0.086 (p=0.505)	CK V54	62	0.121 (p=0.348)
TLC V0	64	0.043 (p=0.738)	TLC V54	64	0.158 (p=0.213)
FVC V0	65	-0.025 (p=0.844)	FVC V54	64	0.005 (p=0.971)
FEV1 V0	65	0.043 (p=0.732)	FEV1 V54	64	0.107 (p=0.401)
DLCO V0	65	0.134 (p=0.288)	DLCO V54	64	0.173 (p=0.172)
Peak TRV V0	59	-0.133 (p=0.315)	Peak TRV V54	54	-0.410 (p=0.002)

Suppl. Table VB	N	correlation coefficient r or ρ^* (p)		N	correlation coefficient r or ρ^* (p)
Length V0	60	0.259 (p=0.046)	Weight V0	60	-0.036 (p=0.786)
Dis dur Rayn V0	59	-0.166 (p=0.210)	Dis dur nonRayn V0	48	-0.190 (p=0.196)
6MWD V0	60	0.722 (p<0.001)	Age V0	60	-0.398 (p=0.002)
mRSS V0	60	-0.092 (p=0.484)	mRSS V54	58	-0.019 (p=0.888)
ESR V0	60	-0.154 (p=0.241)	ERS V54	60	-0.066 (p=0.616)
CRP V0	60	0.051 (p=0.696)	CRP V54	60	-0.136 (p=0.301)
HgB V0	60	0.315 (p=0.014)	HgB V54	60	0.282 (p=0.029)
Creat V0	60	-0.117 (p=0.373)	Creat V54	60	-0.018 (p=0.894)
CK V0	58	-0.185 (p=0.165)	CK V54	57	0.113 (p=0.403)
TLC V0	59	-0.020 (p=0.882)	TLC V54	59	0.227 (p=0.084)
FVC V0	60	-0.101 (p=0.441)	FVC V54	59	-0.025 (p=0.853)
FEV1 V0	60	-0.029 (p=0.823)	FEV1 V54	59	0.065 (p=0.625)
DLCO V0	60	0.123 (p=0.349)	DLCO V54	59	0.258 (p=0.049)
Peak TRV V0	54	-0.098 (p=0.480)	Peak TRV V54	50	-0.460 (p=0.001)

6MWD: six-minute walk distance; V0: baseline visit; Vx: x-month visit; N: number of patients; n: value; Dis dur Rayn: disease duration since first Raynaud; Dis dur non; Rayn: disease duration since first non-Raynauds phenomenon; age; mRSS: modified Rodnan skin score; ERS: erythrocyte sedimentation rate; CRP: C-reactive protein; HgB: haemoglobin; Creat: serum creatinine; CK: creatin kinase; TLC: total lung capacity; FVC: forced vital capacity; FEV1: forced expiratory volume in 1 second; DLCO: diffusing capacity of the lung for carbon monoxide; peak TVR: peak tricuspid valve regurgitation velocity; r: Pearson's correlation coefficient; ρ : Spearman's correlation coefficient. The Pearson's correlation coefficient (r) was used for all variables, except for *, where the Spearman's correlation coefficient (ρ) was used due to skewed distribution.