

Supplementary Table 4. Results of initial and annual evaluations in non-ambulant participants.

		pre	1 year	2 year	3 year	4 year	5 year
Physical evaluation	Summed MMT	24.5 ± 16.2 (20)	23.4 ± 15.2** (18)	23.6 ± 13.1** (15)	17.3 ± 11.2 (13)	19.3 ± 11.3 (9)	18.4 ± 11.2** (13)
	GMFM (%)	22.4 ± 23.3 (17)	19.2 ± 20.4 (16)	15.6 ± 16.2 (14)	8.8 ± 8.4 (11)	7.2 ± 5.6 (8)	7.0 ± 6.8 (11)
	HHD (N)	101.5 ± 74.5 (10)	91.2 ± 91.1 (10)	86.8 ± 103.1 (7)	18.7 ± 25.1 (5)	22.6 ± 26.1 (4)	36.7 ± 65.1 (5)
	Grip power (kg)	3.1 ± 2.9 (10)	2.9 ± 2.7 (10)	2.3 ± 2.3 (10)	2.3 ± 2.2 (7)	1.8 ± 1.7 (6)	1.8 ± 2.2 (8)
	Pinch power (N)	15.5 ± 18.7 (15)	10.8 ± 13.4 (14)	8.2 ± 10.4* (14)	7.7 ± 11.2* (10)	6.3 ± 9.4 (8)	6.1 ± 8.7* (11)
Respiratory function	FVC (%)	75.4 ± 23.3 (20)	71.0 ± 24.3* (19)	76.1 ± 20.5* (15)	66.6 ± 21.6* (13)	65.2 ± 17.1* (8)	62.3 ± 27.6* (15)
	VC (%)	75.2 ± 22.2 (20)	72.4 ± 24.4 (19)	78.2 ± 19.5 (15)	69.5 ± 19.0 (13)	67.1 ± 15.6* (8)	64.3 ± 26.0* (15)
DEXA	Whole-body lean body mass (kg)	28.8 ± 7.1 (20)	28.6 ± 7.8 (18)	28.4 ± 8.3 (14)	27.6 ± 6.2 (14)	25.2 ± 5.3 (9)	26.6 ± 5.8 (14)
	Arm lean body mass (kg)	2.4 ± 0.9 (20)	2.4 ± 1.0 (18)	2.4 ± 1.0 (14)	2.5 ± 0.9 (14)	2.2 ± 0.6 (9)	2.3 ± 0.9 (14)
	Leg lean body mass (kg)	7.5 ± 2.4 (20)	7.7 ± 2.7 (18)	7.7 ± 2.9 (14)	7.5 ± 2.1 (14)	6.8 ± 1.9 (9)	6.8 ± 1.6 (14)
ADL score	CK (IU/L)	138.4 ± 133.3 (20)	155.4 ± 167.6 (18)	103.7 ± 87.8* (15)	63.2 ± 57.7 (12)	68.6 ± 62.7 (9)	70.4 ± 80.9 (15)
	Barthel index	28.0 ± 25.6 (20)	27.9 ± 24.6 (19)	32.9 ± 28.5 (14)	11.0 ± 2.2 (5)	21.1 ± 11.9 (9)	19.6 ± 16.0 (14)
	mRS	4.3 ± 0.8 (20)	4.1 ± 0.7 (19)	4.2 ± 0.6 (15)	4.4 ± 0.5 (7)	4.1 ± 0.6 (9)	4.3 ± 0.5 (15)
QOL score	SF36	Physical functioning	-12.2 ± 5.0 (20)	-13.2 ± 2.8 (18)	-12.9 ± 3.5 (10)	-13.8 ± 1.5 (6)	-14.4 ± 0.0 (6)
		Role physical	33.0 ± 22.5 (20)	29.5 ± 21.2 (18)	25.8 ± 22.6 (10)	26.4 ± 22.7 (6)	12.0 ± 13.3 (6)
		Bodily pain	44.2 ± 12.3 (20)	41.1 ± 10.2 (18)	47.4 ± 9.7 (10)	37.5 ± 8.8 (6)	43.0 ± 9.9 (6)
		Social functioning	37.7 ± 15.9 (20)	37.0 ± 16.2 (18)	34.5 ± 16.7 (10)	24.8 ± 10.8 (6)	28.0 ± 19.0 (6)
		General health perceptions	42.0 ± 8.6 (20)	40.1 ± 10.1 (18)	40.7 ± 6.3 (10)	38.4 ± 4.0 (6)	38.5 ± 6.7 (6)
		Vitality	45.5 ± 8.9 (20)	40.2 ± 9.9* (18)	42.8 ± 8.3 (10)	40.2 ± 3.5 (6)	39.7 ± 7.4 (6)
		Role emotional	42.3 ± 17.8 (20)	37.4 ± 19.7 (18)	38.2 ± 20.7 (10)	42.2 ± 8.6 (6)	36.0 ± 23.6 (6)
		Mental health	47.7 ± 8.8 (20)	44.7 ± 8.6 (18)	46.2 ± 10.5 (10)	42.0 ± 4.4 (6)	43.3 ± 4.3 (6)
							46.5 ± 10.1 (12)

P-values are calculated relative to baseline data. *: p<0.05, **: p<0.01