

		Baseline	1 year	2 year	3 year	4 year	5 year	
Physical evaluation	Summed MMT	38.6 ± 22.5 (33)	36.5 ± 21.7** (29)	34.7 ± 20.3** (23)	31.7 ± 21.4** (21)	29.7 ± 19.0** (13)	32.3 ± 22.1** (21)	
	GMFM (%)	50.4 ± 38.4 (29)	46.3 ± 38.7 (26)	42.6 ± 39.0* (22)	41.0 ± 39.9* (19)	28.6 ± 37.1 (11)	33.9 ± 37.1** (18)	
	6MWT	350.5 ± 144.4 (12)	307.5 ± 134.1 (11)	307.0 ± 147.8 (8)	265.5 ± 102.9 (8)	222.0 ± 130.1** (4)	270.8 ± 112.2 (4)	
	HHD (N)	163.1 ± 113.0 (23)	152.0 ± 145.6 (21)	132.0 ± 97.0 (15)	144.4 ± 137.1 (13)	116.3 ± 161.4 (8)	130.9 ± 146.1 (12)	
	Grip power (kg)	9.1 ± 8.1 (23)	6.3 ± 6.4 (20)	6.4 ± 7.6 (18)	7.2 ± 7.5 (15)	4.6 ± 5.6* (10)	6.1 ± 8.2* (16)	
Respiratory function	Pinch power (N)	26.5 ± 22.2 (27)	20.6 ± 20.3 (24)	19.7 ± 21.4** (22)	15.0 ± 15.0** (18)	12.1 ± 12.9* (12)	16.3 ± 20.2** (19)	
	FVC (%)	87.0 ± 25.0 (33)	84.3 ± 27.6 (30)	87.4 ± 25.0* (23)	81.2 ± 26.8** (22)	75.0 ± 24.0** (12)	75.8 ± 31.2** (23)	
	VC (%)	86.9 ± 24.2 (33)	85.5 ± 27.3 (30)	88.9 ± 23.4 (23)	83.3 ± 24.8* (22)	76.5 ± 21.5** (12)	77.9 ± 29.7** (23)	
DEXA	Whole-body lean body mass (kg)	31.1 ± 7.1 (32)	30.6 ± 7.2 (29)	30.3 ± 7.3 (23)	30.0 ± 6.5 (22)	27.7 ± 5.8 (14)	28.7 ± 6.2 (20)	
	Arm lean body mass (kg)	2.7 ± 1.1 (32)	2.7 ± 1.1 (29)	2.6 ± 0.9 (23)	2.7 ± 1.0 (22)	2.5 ± 0.8 (14)	2.6 ± 0.9 (21)	
	Leg lean body mass (kg)	8.6 ± 2.6 (32)	8.5 ± 2.5 (29)	8.5 ± 2.6 (23)	8.3 ± 2.2 (22)	7.3 ± 1.7 (14)	7.7 ± 2.1 (21)	
CK (IU/L)		268.3 ± 290.8 (33)	270.1 ± 336.9 (29)	203.0 ± 228.5 (24)	186.0 ± 227.8* (21)	177.3 ± 208.4* (13)	151.5 ± 172.6* (23)	
ADL score	Barthel Index	54.8 ± 39.6 (33)	50.4 ± 38.9 (28)	54.5 ± 39.0 (21)	64.6 ± 44.2 (13)	38.8 ± 33.8 (12)	42.9 ± 36.8 (21)	
	mRS	3.6 ± 1.2 (33)	3.5 ± 1.1 (28)	3.6 ± 1.1 (22)	3.3 ± 1.2 (15)	3.9 ± 0.7 (12)	3.9 ± 1.1 (22)	
QOL score	SF36	Physical functioning	0.0 ± 19.3 (33)	-4.9 ± 15.4 (27)	1.2 ± 21.3 (16)	0.8 ± 16.3* (14)	-2.5 ± 13.9* (14)	-5.1 ± 16.8* (21)
		Role physical	35.4 ± 19.1 (33)	30.7 ± 17.9 (27)	30.6 ± 20.5 (16)	31.8 ± 16.0 (14)	25.1 ± 18.2 (14)	27.1 ± 18.8 (21)
		Bodily pain	45.1 ± 11.3 (33)	42.0 ± 9.7 (27)	47.6 ± 9.7 (16)	42.5 ± 11.1 (14)	43.4 ± 10.7 (14)	41.5 ± 11.7 (21)
		Social functioning	38.1 ± 13.9 (33)	38.6 ± 15.0 (27)	35.7 ± 15.0 (16)	30.3 ± 9.4 (14)	31.7 ± 19.1 (14)	34.9 ± 17.3 (21)
		General health perceptions	40.0 ± 9.3 (33)	39.2 ± 9.1 (27)	40.8 ± 6.8 (16)	37.0 ± 7.1 (14)	35.8 ± 9.7 (14)	37.1 ± 9.0 (21)
		Vitality	44.5 ± 10.8 (33)	42.0 ± 10.2 (27)	43.8 ± 10.5 (16)	41.4 ± 8.7 (14)	39.5 ± 11.1 (14)	39.6 ± 14.8 (21)
		Role emotional	41.8 ± 16.6 (33)	40.4 ± 18.0 (27)	41.3 ± 18.9 (16)	46.3 ± 8.9 (14)	38.2 ± 18.9 (14)	34.3 ± 19.9 (21)
		Mental health	46.0 ± 11.2 (33)	47.2 ± 8.9 (27)	47.5 ± 10.2 (16)	46.7 ± 7.8 (14)	43.6 ± 10.3 (14)	43.0 ± 12.6 (21)