

**Supplementary table 1. Frequency of clinical features**

Clinical feature	Number of patients (n=10)
Exercise intolerance	10
Sensorineural hearing loss	9
Ataxia	8
Constipation	5
Diabetes mellitus	5
Fatigue	5
Migraine	5
Muscle weakness	5
Depression	4
Epilepsy	2
Myalgia	2
Ophthalmoparesis	2
Ptosis	2
Retinopathy	2
Underweight (BMI <18.5)	2
Cognitive decline	1
Dysarthria	1
Dysphagia	1
Encephalopathy	1
Hypertension	1
Hypothyroidism	1
Neuropathy	1
Short stature	1

BMI = body mass index

**Supplementary table 2. Body composition and blood analyses before and after training**

Parameter	Patients			Controls			Interaction <i>p</i> value <sup>b</sup>
	Baseline	Follow up	<i>p</i> value <sup>a</sup>	Baseline	Follow up	<i>p</i> value <sup>a</sup>	
Body composition							
Weight (kg)	62.1 ± 12.2	62.8 ± 11.8	0.150	81.9 ± 12.9**	81.6 ± 12.8**	0.777	0.909
BMI	21.3 ± 3.4	21.5 ± 3.3	0.154	27.7 ± 4.7**	27.6 ± 4.4**	0.670	0.907
Lean body weight (kg)	46.6 ± 12.7	47.6 ± 12.4	0.056	55.7 ± 9.2	55.8 ± 8.8	0.728	0.944
Fat (% of body weight)	25.7 ± 8.9	25.1 ± 7.6	0.509	31.7 ± 11.1	31.0 ± 10.0	0.421	0.993
Blood analyses							
Lactate	2.1 ± 0.5	2.6 ± 0.9	0.132	2.3 ± 1.0	1.9 ± 0.8	0.648	0.202
Creatine kinase	121 ± 46	143 ± 77	0.404	125 ± 49	124 ± 55	0.958	0.833

BMI = body mass index; <sup>a</sup> *p* value represents the within group comparison of paired before and follow up time points; <sup>b</sup> *p* value represents group by time interaction from mixed model repeated measures analysis; \* *p*<0.05 and \*\* *p*<0.01 for patients vs controls at equivalent time points

**Supplementary table 3. Disease burden and quality of life before and after exercise training**

<b>Score</b>	<b>Baseline</b>	<b>Follow-up</b>	<b><i>p</i> value</b>
NMDAS	16 ± 7	15 ± 7	0.269
SF-12 MHC	50 ± 9	48 ± 14	0.673
SF-12 PHC	45 ± 12	44 ± 12	0.434

NMDAS = Newcastle Mitochondrial Disease Adult Scale; FIS = Fatigue Impact Scale; SF-12 = Short Form 12 Health Questionnaire; MHC = mental health component; PHC = physical health component.