

Table S3. Increased respiratory chain activities in skeletal muscle of the patient and similar respiratory chain activities in myoblasts of the patient and control, in basal and pro-inflammatory conditions. T+I : TNF- α + IL-1 β .

Frozen skeletal muscle	Control		Patient	
Spectrophotometric activities (nmol/min/mg proteins)				
Complex I	17 \pm 4		56	
Complex II	35 \pm 7		105	
Complex III	300 \pm 56		600	
Complex IV	150 \pm 30		300	
Citrate synthase	96 \pm 18		228	
Lactate dehydrogenase	1758 \pm 415		3800	
Myoblasts	Control		Patient	
Spectrophotometric activities (nmol/min/mg proteins)	Base	T+I	Base	T+I
Complex I	13.8	11.4	10.5	10.4
Complex II	15.5	13.7	11.9	11.2
Complex III	168.4	176.7	156.8	147.1
Complex IV	46.3	42.8	35.7	35.3
Citrate synthase	51.8	51.2	44.6	44.8
Lactate dehydrogenase	9276.7	9875.3	8538.3	8882.7