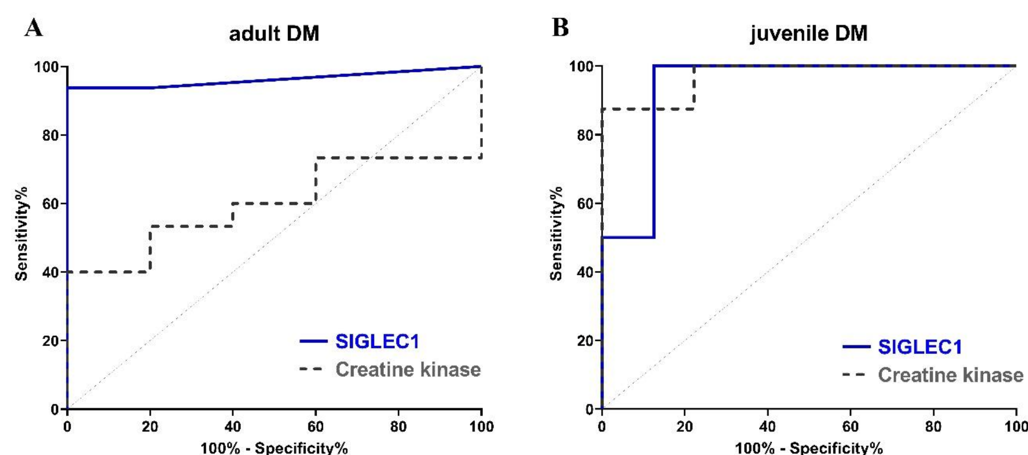
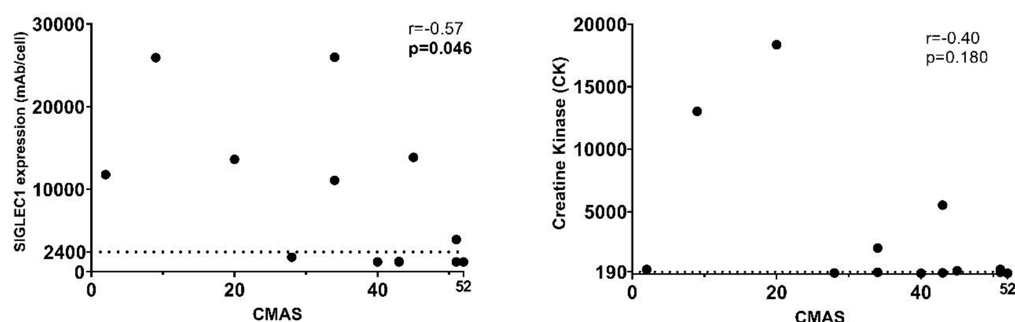
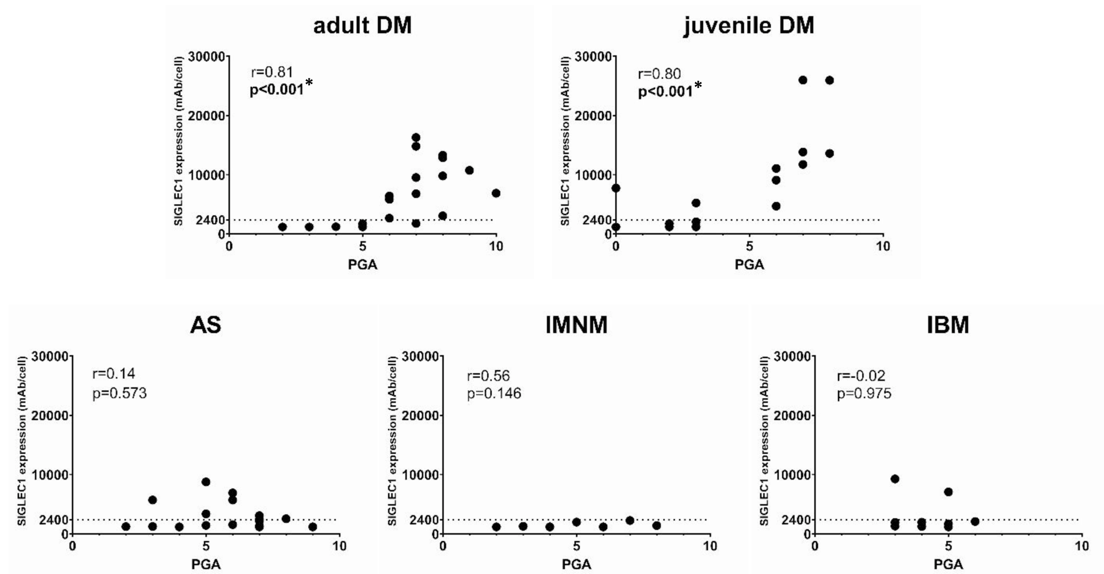
Supplemental material



eFigure S1 Receiver operating characteristic curves for SIGLEC1 and creatine kinase in dermatomyositis (DM) patients. The curves show the ability of each biomarker to distinguish between patients with PGA≥5 (moderate to severe disease activity) and PGA<5 (no to moderate disease activity), in **(A)** adult DM patients (n=21); SIGLEC1: AUC=0.96, p=0.002; creatine kinase: AUC=0.60, p=0.513 and **(B)** juvenile DM patients (n=17); SIGLEC1: AUC=0.97, p=0.001; creatine kinase: AUC=0.94, p=0.003



eFigure S2 Correlation between biomarkers and muscle strength scores (CMAS) in juvenile dermatomyositis patients at assessment visit 1 (VC1) (A) SIGLEC1 versus CMAS (n=13) and (B) creatine kinase versus CMAS (n=13); Asterisks (*) represent significant results (p<0.05). Spearman's rank test was used for both analyses; VC1, first visit with assessment of SIGLEC1 and CMAS

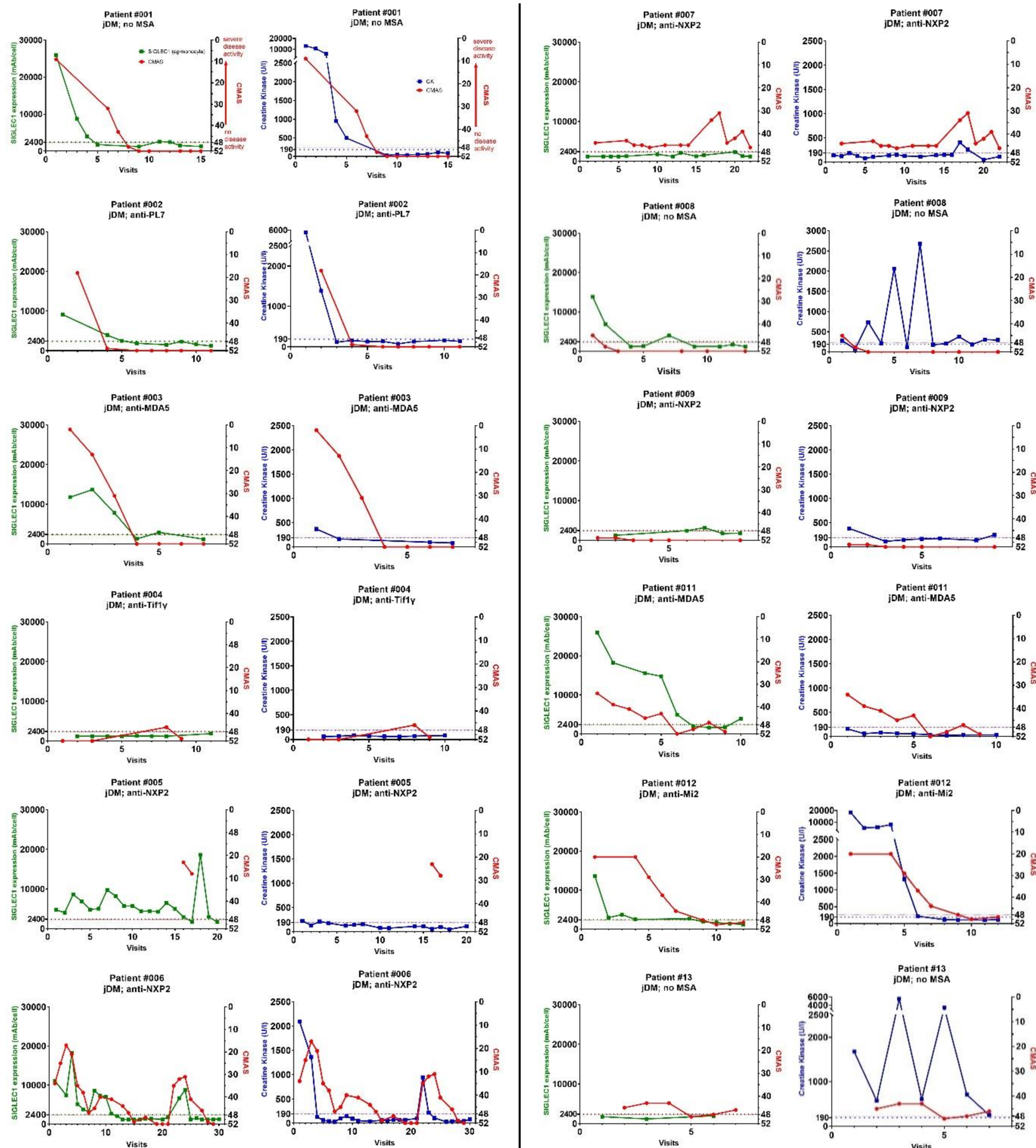


eFigure S3 Correlation between SIGLEC1 expression on monocytes and Physician Global Assessment (PGA) at first visit for the various IIM subgroups. Asterisks (*) represent significant results (p<0.05). Spearman's rank test was used for all analyses.

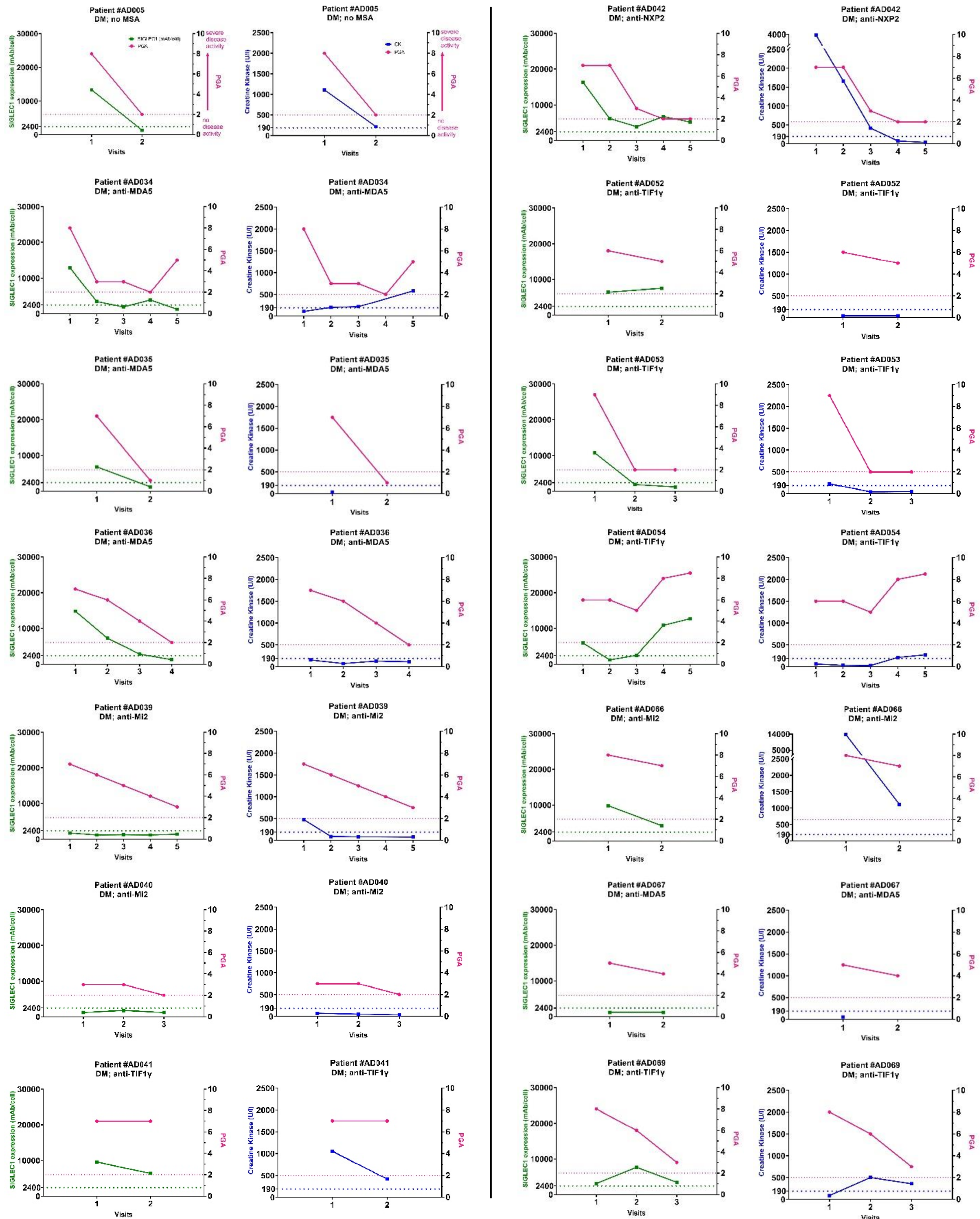
Patient ID	Clinical diagnosis	Myositis-specific antibodies (MSA)	Myositis-associated antibodies (MAA)	SIGLEC1, mAb/cell (blood)	MxA status (muscle)	ISG15 status (muscle)
AD029	AS	Jo1	Ro52	5721	0	1
AD031	AS	Jo1	Ro52	2568	0	0
AD030	AS	Jo1	Ro52, PM-Scl100	2449	0	1
AD039	DM (adult)	Mi2	Ro52	1769	0	0
AD042	DM (adult)	NXP2	-	16295	1	1
AD005	DM (adult)	-	Ro52	13301	0	0
AD041	DM (adult)	TIF1γ	-	9545	0	0

AD067	DM (adult)	MDA5	PM-Scl100	1200	0	0
AD065	DM (adult)	MDA5	-	6901	0	0
PAE002	DM (juvenile)	PL7	-	9114	0	1
PAE001	DM (juvenile)	-	-	25931	1	1
AD010	IBM	-	Ro52, Ku, U1RNP	9281	1	0
AD004	Overlap	-	Ku	8305	0	1
AD072	IMNM	HMGCR	-	1752	0	0
AD008	Overlap	-	Ro52, Ku, PM-Scl75, PM-Scl100	1645	0	0
AD047	Overlap	SRP	-	1260	0	0
AD064	Overlap	-	Ku	1200	0	0

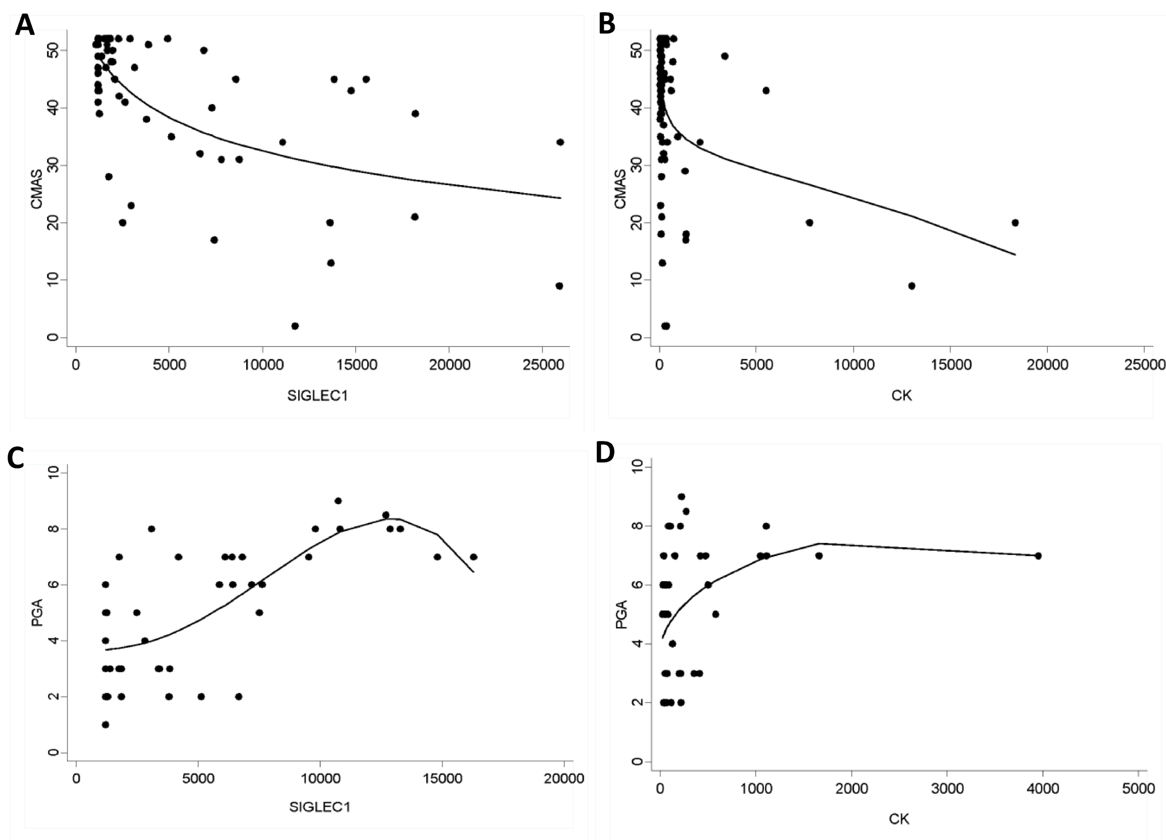
eTable S1 Subgroup analysis of SIGLEC1 expression (in blood) versus MxA and ISG15 status (in immunohistochemical muscle biopsy stains) (n=17). MxA/ISG15 status: 0 = negative, 1 = positive



eFigure S4 Longitudinal graphs of SIGLEC1 vs. CMAS and creatine kinase vs. CMAS for all juvenile DM patients with at least two visits; PGA, Physician Global Assessment; CMAS, Childhood Myositis Assessment Scale



eFigure S5 Longitudinal graphs of SIGLEC1 vs. PGA and creatine kinase vs. PGA for all adult DM patients with at least two visits; PGA, Physician Global Assessment; CMAS, Childhood Myositis Assessment Scale

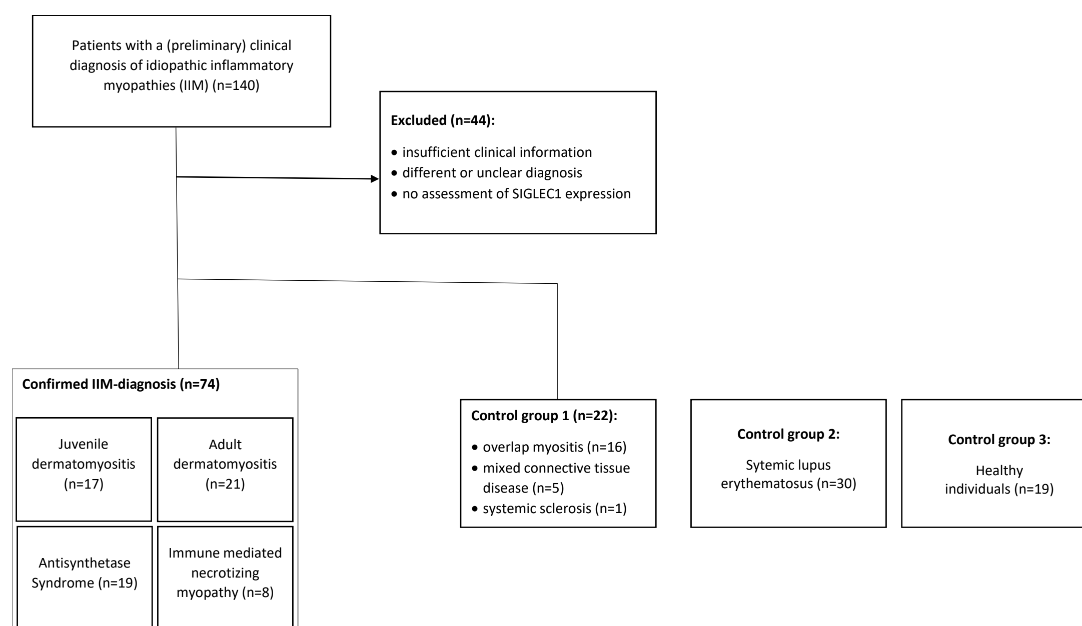


eFigure S6 Visualization of the results of longitudinal data analysis (see Table 2) of A) SIGLEC1 vs. CMAS for juvenile DM patients (absolute values); B) Creatine kinase (CK) vs. CMAS for juvenile DM patients (absolute values); C) SIGLEC1 vs. PGA for adult DM patients (absolute values); D) Creatine kinase (CK) vs. PGA for adult DM patients (absolute values)

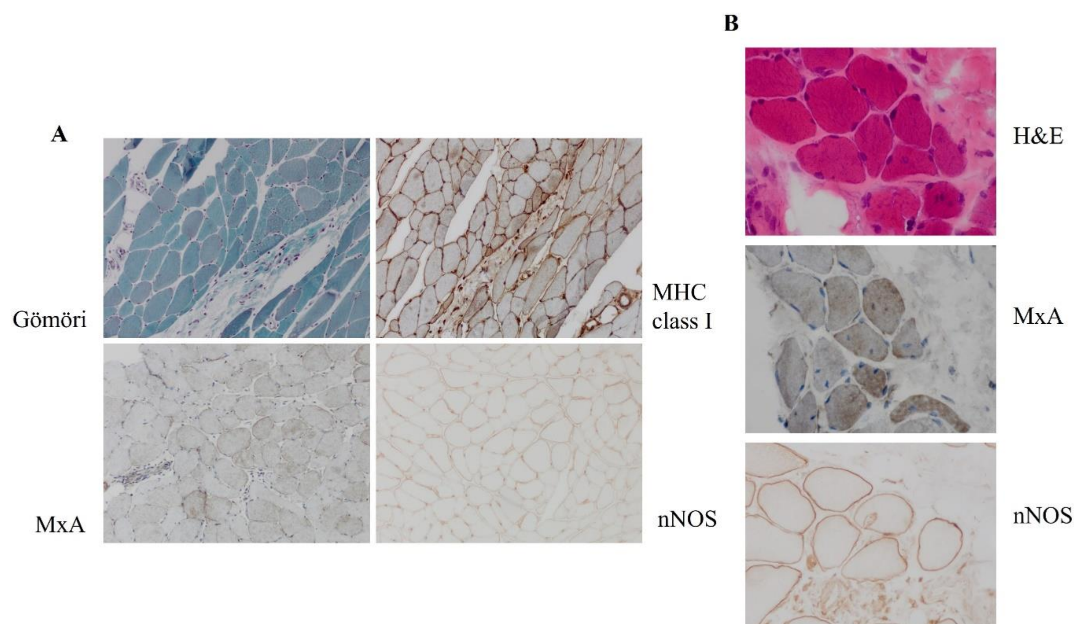
eText S1: Laboratory measurement of SIGLEC1

SIGLEC1 expression was determined by flow cytometry using a highly standardized quantitative assay. Briefly, 25µl of EDTA-anticoagulated whole blood was incubated with 10 µl of mouse-anti-human antibody cocktail containing phycoerythrin (PE)-labeled anti-CD169 monoclonal antibody (mAb) (labeled at a fluorochrome/protein ratio of 1:1), allophycocyanin (APC)-labeled anti-CD14 mAb and KromeOrange-labeled anti-CD45 mAb for 15 min at room temperature (RT) in the dark (all antibodies from Beckman Coulter, Krefeld, Germany). Red blood cells were then lysed by adding 500µl of Versa-Lysis solution (Beckman Coulter) to each reaction tube. After incubation for 30 min at RT in the dark,

samples were centrifuged for 5 min at 200 x g at RT. Samples were then washed once with 1000 µl PBS containing 2% fetal calf serum (FCS) and centrifuged again for 5 min at 200 x g at RT. Stained samples were acquired on a 10-color flow cytometer (Navios, Beckman Coulter) and analyzed using the Navios software. During each analytical run, QuantiBRITE™ PE tubes (BD Biosciences) were acquired to convert the fluorescent channel 2 (FL2) mean fluorescent intensity (MFI) signals on $CD14^+$ monocytes to monoclonal antibodies bound per cell (mAb/cell) values. FL2 MFI values and absolute values for PE molecules (as specified by the manufacturer) for each QuantiBRITE™ bead population were used to perform linear least square regression analysis to determine the best calibration value, which then was used to convert the FL2 MFI values of monocytes in the analytical sample to the amount of PE-labeled CD169 mAb bound per monocyte (mAb/cell).



eFigure S7 Flow diagram of study participants



eFigure S8 A-B Negative control stainings with an irrelevant antibody for MxA: As a negative control we used the species specific antibody (rabbit polyclonal) nNOS (neuronal nitric oxide synthase) at the same dilution (1:100). This is an antibody that we use in every muscle biopsy work-up to demonstrate integrity of the sarcolemma or lack of staining in e.g. denervation processes. The macrophages, endothelial cells and the sarcoplasm are not stained by nNOS. nNOS can similarly be used as a negative control with the species specific antibody (rabbit polyclonal) for ISG15 at a dilution 1:50.