Supplementary Table S1. Summary of main results of studies exploring IIMs pathogenesis.

Population (n)	Main results	Author
PM (n=363) DM (n=654) (Chinese population)	- Only rs3733197 resulted significantly associated to DM +PM - Only rs3733197 resulted significantly associated with DM and PM/DM patients with ILD involvement	Chen *et al*. (1)
PM (n=88) and DM (n=63) *vs*. HC (n=116) Vietnamese pts	HLA- DRB*13 was significantly associated to risk of PM development but not to DM. HLA-DRB1*09 was protective for DM.	Nguyen *et al*. (2)
IIMs (n=74) pts (58 DM, 16 PM) HC (n=22), SLE pts (n=20), SSc pts (n=20), RA pts (n=20)	- PMN elastase was significantly higher in IIMs compared to HC - PMN elastase was significantly higher in IIMs with high disease activity	Wu S. *et al*. (4)
IIMs complicated by ILD (n=30) *vs*. HC (n=30) *vs*. pulmonary infection (n=30)	- Serum MCP-1 and TGF-β1 levels were significantly higher in PM/DM pts compared to HC and to pulmonary infection group.	Wu C.Y. *et al*. (5)
IIMs (n=110: 40 DM, 40 PM, 30 CAM) *vs*. HC (n=42).	- S100A11 was detectable in cytoplasm of necrotising and regenerating myofibres. - Plasmatic S100A11 was significantly higher IIMs compared to HC	Cerezo *et al*. (6)
IIMs (n=148; 109 DM, 39 PM, 28 anti-Jo1 positive, 52 anti-MDA5 positive and 68 antibodies negative) *vs*. 56 HC	- MMP9 mRNA levels in PBMCs was significant higher in IIMs pts compared to HC - MMP9 mRNA levels are similar in anti-Jo1, anti-MDA5 and antibodies negative pts compared to HC - MMP9 serum level was not significantly different between IIMs and and between patient groups with and without ILD - MMP9 serum level was significantly higher in anti-Jo1 positive pts compared to anti-MDA5 positive, antibody-negative , and HC -In anti Jo1 positive MMP9 was more released from isolated neutrophils than from isolated PBMCs	Liu *et al*. (7)
IIMs (n=119: 39 DM, 49 IMNM, 18 ASSD, 13 IBM) *vs*. HC (n=20)	- The IFN1 inducible gene are markedly elevated in DM, moderately expressed in ASSD and minimally expressed in IBM and IMNM. - The IFN2 specific genes are increased in IBM, ASSD, and DM biopsies compared to HC. - IFN2 inducible genes are much lower in IMNM compared to HC and other IIM (DM; IBM and ASS) - IFN1 and IFN2inducible gene had a positive correlation with gene associated to inflammatory cells (T-cells and macrophages) and to muscle regeneration - Higher level of IFN inducible gene correlate with higher CK level and decreased muscle strength	Pinal-Fernandez *et al*. (8)
IIMs (n=47: 32 DM, 13 PM and 3 ASSD)	- DM patients had significantly higher IFN1 signatures compared to other IIMs. - MDA5 positive pts had highest IFN1 signature compared to ARS group and double negative pts - No significative difference was found in pts with ILD.	Ono *et al*. (9)
DM patients with anti-NXP-2 (n=13) *vs*. DM with anti-MDA5 (n=6), DM with anti-Mi2 (n=7) and HC (n=11).	- Anti-NPX-2 pts showed vasculopathy of perimysial arterioles - MxA was expressed in endothelial cells, smooth muscle cells and pericytes of the diseased arterioles. - VEGF was low in endomysial neovessels	Liu *et al*. (11)
IMNM (n=10), DM (n=10), ASSD (n=10), IBM (n=9) and HC (n=10)	- ISG15 expression was higher in muscle sample from DM pts compared to IMNM, ASSD and IBM - Gene expression induced by IFNγ was high in ASSD and IBM, low in DM and absent in IMNM: 1) Immunohistochemical distribution of IFNγ inducible MHC-2 was perifascicular in ASSD and multifocal in IBM. 2) CIITA gene was higher in ASSD and IBM compared with IMNM and DM 3) HLA-DOB was significantly higher in IBM and in ASSD 4) HLA-DPB was significantly higher in IBM and ASSD both compared to IMNM. 5) HLA-DRB genes was significantly higher in IBM compared with IMNM 6) HLA-DPB gene was significantly higher in IBM and ASSD borth compared to DM. 7) GBP2 gene was significantly higher in IBM and ASSD both compared to IMNM. 8) GBP2 gene was significantly higher in ASSD	Rigolet *et al*. (10)
71 pts (PM n= 10 and DM n=61) *vs*. HC (n=30)	- CD3+T and CD4+T are markedly decreased in PM and DM compared to HC. - In DM CD8+T and NK cells are significantly reduced compared to HC. - B cells are significantly reduced in PM compared to HC. - Treg cells and Th1 cells were significant lower both in PM and DM compared to HC - Th17/Treg in PM/DM are significantly higher than that in HCs - sIL-2Rα is higher in PM/DM compared to HC - IL-2 level in PM/DM is significantly lower than HC - Other cytokines are higher in PM/DM than HC: IL-4 (only in DM), IL-6, IL-10, TNF-α (only in DM), IFN-γ and IL-17 - In DM pts treated with Id-IL-2 the number of Tregs is significantly upregulated. - Disease activity (MYOACT) was negatively correlated to T cell, IL-2 levels, ESR, CRP and positively correlated to B cells percentage and muscle enzymes level. - Tregs deficiency was an independent risk factor for ILD	Feng *et al*. (13)
44 pts (11 PM and 33 DM) *vs*. 20 HC	- mTRAIL signal was weak in HC and strong in muscle tissue from PM and DM pts (primarily located in the infiltrated mononuclear cells) - Serum TRAIL (s TRAIL) was significantly higher in PM pts and DM pts compared to HC - mTRAIL and its receptors, including DR4 and DR5, was significantly elevated on circulating T cells from PM and DM pts than HC. - sTRAIL positively correlate with disease activity scoring by MYOACT tool, with dysphagia and Jo1 positivity. - sTRAIL decrease after 3 to 6 months of therapy.	Zhou *et al*. (14)

Population (n)	Main results	Author
IIMs pts (n=47) *vs*. HC (n=30)	- CD4+CXCR5+CCR7^{lo}PD-1^{hi} T cells was significantly increased in IIMs compared to HC - High levels of CD4+CXCR5+CCR7^{lo}PD-1^{hi} T cells was found in pts with anti-NPX2 positivity antibodies, while low levels were found in pts who were positive for anti-TIFγ and anti-HMGCR - Intracellular IL-21 expression was significantly more frequent in CD4+CXCR5+CCR7loPD-1hi T cells from IIMs pts compared to HC - IL-21 and CD4+CXCR5+CCR7loPD-1hi T cells positively correlate with disease activity and reduced after 6 months of therapy	Zhang*et al*. (15)
IIMs-ILD (n= 85: 17 PM-ILD, 25 DM-ILD and 43 ADM-ILD); IIMs without ILD (n= 19: 4 PM, 8 DM and 7 ADM); patients with RA-ILD (n=14), patients with SSc-ILD (n=7), patients with idiopathic pulmonary fibrosis (n= 6), patients with community acquired pneumonia (n=7), and HC (n=13)	- CD4+CXCR4+ T cells is increased among PBMCs and BALF in IIMs-ILD patients compared with disease controls and HC. -Serum KL-6 concentrations were also elevated in IIMs-ILD patients compared with HC but it should be not considered specific, being observed also in disease controls. - SDF-1 was elevated in the serum of IIMs-ILD patients compared with other controls - The peripheral percentage of CD4+CXCR4+ T cells was significantly correlated with the extent of structural changes, as identified by HRCT score and reduction of FVC and DLCO - Pts with high CD4+CXCR4+ (>30%) tend to have negative prognostic factors as baseline high HRCT score, low FVC%, higher ferritin levels, presence of anti-MDA5 and ADM phenotype. - CD4+CXCR4+ T cells (>30%) was an independent risk factors for mortality and decrease among survivors within the first month after treatments	Wang *et al*. (16)
IIMs/ASSD pts (n=24) *vs*. sarcoidosis pts (n=7) and HC (n=12)	In BALF: - CD4+T response to HisRS- protein was found in 3/4 pts; response to HisRS11-23 was found 2/3 IIMs/ASSD (median CD40L fold-change: 88; 27-149). In PBMCs: - CD4+ T response to HisRS-protein was found in 14/18 IIMs/ASSD; response to HisRS11-23 was found in 11/14 IIMs/ASSD - CD4+T response to HisRS11-23 was found in 3/7 sarcoidosis, and in 5/12 HCs - Number of CD4+CD40L+ T cells that produced in FNγ was significantly higher from BALF compared to cells isolated from blood both after stimulation of HisRS and HisRS11-23 - Anti-Jo1 autoantibodies were detected in BALF in 6/7 anti-Jo1+ patients, in 2/9 sarcoidosis and not detected in anti-Jo1- pts and HC	Galindo-Feria *et al*. (17)
DM (n=13) *vs*. HC (n=18)	- DM pts had higher proportion of naïve CD4+Tcells and lower CD4+ TEM-cell than HC. - DM pts had lower proportion of naïve CD8+ T and CD8+ TCM-cell. - Treg-cell, Th1 and Tfh-cell proportions were lower in pts. - Th2 and Th17 are similar between DM and HC. - DM pts had higher proportion of naïve B-cell and lower proportion of memory B-cell. - Presence of ILD contributed to the differences between DM and HC. - After treatment there was no change in proportion of T-cells while there was an increase of memory B-cell.	Sasaki *et al*. (18)
IBM (n= 40); other IIMs (n=77); mitochondrial non-inflammatory myopathy (n=188); HC (n=106).	- Muscle gene expression demonstrate an increased cytotoxic lymphocyte signature in IBM distinct from all other muscle diseases. - Abundance of T cell-related chemokines and cytokines, in particular IFNG and its induced chemokines and cytokines CXCL9, CCL5, and IL32 was present -High differentiated CD8+cytotoxic T cell are high represented in IBM muscle (more than in PM)	Greenberg *et al*. (19)
-IBM n=10 in cohort 1 and n=8 in cohort 2. -16 HC (8 subject matched for age and sex with each IBM cohort). -ASSD Jo1 positive n =10, -anti-HMGCR IMNM, n=9 - anti-SRP IMNM, n=7	- CD8T-bet+ are significantly more frequent in IBM patients compared to healthy donors. - CD8+ terminally differentiated memory T cells ((CD8+ CD45RA+ CCR7) are significantly more frequent in IBM pts compared to HC - Marked increase in the expression of CD57 within CD8T-bet+ cells in IBM pts compared to HC, in both CD8T-bet+ cells and CD8T-bet- cells - CD8T-bet+ CD57+ cells are CD28null CD27null CD127null in both pts and HC - Decrease in expression of CD28, CD27 and CD127 in non-senescent CD8T-bet+ CD57- cells of sIBM pts compared to HC. - CD8T-bet- cells presented CD28, CD27 and CD127 (both in pts and HC)	Dzangue-Tchoupou *et al*. (20)
IBM (n=10) *vs*. HC (n=10)	- TDP-43 was increased in IBM compared to HC and colocalised with mitochondria	Huntley *et al*. (21)
IBM (n=11) *vs*. controls (n=11: 2 PM, 3 DM 1 spinobulbar muscular atrophy, 1 progressive muscular atrophy, 1 amyotrophic lateral sclerosis and 3 HC)	CYLD expression in degenerative myofibres of IBM	Yamashita *et al*. (22)
PM/DM (n=24) *vs*. HC (n=12) Animal models: of EAM (n=6) *vs*. control (n=6)	- CD163 was increased in PM/DM muscle samples while no expressed in HC. - Muscle mRNA levels of MCP-1, TNF-α, IL-6 were increased in pts compared to HC - Muscle mRNA levels of Nef2 was reduced in pts compared to HC - Serum CK, ROS and pro-inflammatory factors (MCP-1, TNF-α, IL-6) were significantly higher in pts compared to HC. Macrophages isolated from EAM transfected by Nrf2 showed: - significant decrease of the mRNA and protein levels of MCP-1, TNF-α, IL-6 - promotion of mRNA and protein levels of total Nrf2 and of antioxidant protein. - significant reduction of migration ability	Liu *et al*. (24)

PM: polymyositis; DM: dermatomyositis; HC: healthy controls; pts: patients; SNP: single nucleotide polymorphism; BANK1B: cell scaffold protein with ankyrin repeats 1 BANK1; IIMs: idiopathic inflammatory myopathies; OR: odds ratio; PMN: polymorphonuclear; ENR: elastase-to-neutrophil ratio; RA: rheumatoid arthritis; SLE: systemic lupus erythematosus; SSc: systemic sclerosis; CAM: cancer associated myositis: MYOACT: myositis disease activity assessment; MMP9: matrix metalloproteinases-9; ASSD: antisynthetase syndrome; IBM: inclusion body myositis; IMNM: immune mediated necrotising myopathy; IFN: interferon; IFN1: type I interferon; IFN2: type II interferon; CK: creatine kinase; MDA5: melanoma differentiation-associated protein 5; ARS: antibodies to T-RNA synthetase; NPX2: nuclear matrix protein 2; BALF: bronchoalveolar lavage fluid; KL-6: Krebs von den Lungen-6; SDF-1: stromal cell derived factor-1; MCP-1: monocyte chemoattractant protein-1; TGF-β1: transforming growth factor-β1; ASSD: anti-synthetase syndrome; HMGCR: 3-hydroxyl-3-methylglutaryl CoA reductase; SRP: signal recognition particle positive; ADM: amyopathic dermatomyositis; PBMCs: peripheral blood mononuclear cells; CyTOF: cytometry by time of flight; MMI: median mass intensities; IL-: interleukin; TNF-α: tumour necrosis factor-α; IFN-γ: interferon-γ; sIL-2Rα: soluble interleukin-2 receptor α; ld-IL2: low-dose IL-2; CYLD: cylindromatosis; MxA: myxovirus resistance protein A; VEGF: vascular endothelial growth factor; EAM: experimental autoimmune myositis; CK: creatine kinase; ROS: reactive oxygen species; MCP-1: mast cell protease-1; TCM: central memory T; TEM: effector memory T; T1-IFN: type 1 interferon.