



Figure 1. Free thiols per gram protein in all groups * $p < 0.005$ between timepoint 2 and 3

Although these findings need further study, they may suggest activation of ubiquitous antioxidant defense mechanisms during cooling and/or rewarming and should be explored for future use as a potential therapeutic target in RP.

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FRI0367 NEW AUTOIMMUNE TARGETS IN IDIOPATHIC INFLAMMATORY MYOPATHIES - AN ANTIGEN BEAD ARRAY APPROACH

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Background: The Idiopathic Inflammatory Myopathies (IIM) is a group of rare systemic inflammatory diseases characterized by severe organ involvement and premature mortality. Several myositis specific auto-antibodies (MSAs) have been recognized and associated with specific clinical manifestations and prognosis, still many patients are autoantibody negative. Identification of new autoimmune targets will be helpful in improving diagnosis, better stratifying into subgroups, prediction of prognosis, tailoring treatment and to understand underlying biological pathways.

Objectives: To identify new autoimmune targets in IIM by antigen bead array (1).

Methods: A bead array with 354 antigens was used to explore the autoimmune reactivity in 881 plasma samples from patients with IIM (N=225), Systemic Lupus Erythematosus (SLE) (N=350) and population controls (N=306). The antigens were selected from initial screenings of 160 SLE-samples on a total of 5760 antigens on planar arrays, and a first verification bead array with 355 antigens. The IIM samples represented three groups of patients with distinct diagnoses: Dermatomyositis (DM, N=83), Polymyositis (PM, N=111) and Inclusion Body Myositis (IBM, N=31), who were regularly followed at the Rheumatology Unit of the Karolinska University Hospital from January 2003 until March 2014. Based on 2 possible levels of cutoff, each sample was classified as reactive to each single antigen (Ag) at low or high cut off or non-reactive.

Results: In general, depending on the cutoff stringency, 86–88% of the 354 selected antigens showed reactivity in at least one sample with no difference between IIM, SLE and controls. Comparing PM, DM, IBM according to the number of samples which showed reactivity towards each single Ag, reactivity at high cut off towards NADH dehydrogenase 1 α subcomplex 11 (NDUFA11), poly(A) RNA polymerase D4 (PAPD4), CD163, I(3)mbt-like 1 (L3MBTL1) and calcium release-activated calcium modulator 2 (ORAI2) was discovered with higher frequencies in the IBM samples compared to PM and DM. In the group of IIM patients testing negative for all the known MSAs increased reactivity at high cut off was observed towards E3 ubiquitin protein ligase 2 (SIAH2), leiomodulin 2 (LMO2) and RAD23 homolog A (RAD23A). In the group of IIM patients with history of malignancy and no evidence for anti-p155/140 antibodies the antigens early B-cell factor 2 (EBF2), POU class 6 homeobox 1 (POUF61) and growth differentiation factor 7 (GDF7) revealed high serum reactivity. In IIM patients with interstitial lung disease increased reactivity at high cut off was found towards zinc finger protein 688 (ZNF688) and prostaglandin D2 receptor (PTGDR). A high frequency of known target reactivities (MSAs) was also confirmed.

Conclusions: Reactivity towards autoantigens corresponding to human proteins was present in plasma samples from IIM, controls, and SLE. Potentially new

autoimmune targets have been discovered in IIM subgroups, although further validation in independent cohorts is needed.

References:

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FRI0368 NAILFOLD CAPILLAROSCOPY CHANGES REFLECT ENDOTHELIAL ACTIVATION AND INJURY IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a severe connective tissue disease characterized by vascular and fibrotic changes in the skin and various internal organs. Pathogenesis of SSc includes early-onset vasculopathy with endothelial cell activation, microvascular injury and impaired angiogenesis.

Objectives: We aimed to determine the association of several biological molecules reflecting endothelial cell activation or dysfunction: E-selectin (E-sel), inter-cellular adhesion molecule 1 (ICAM-1), endothelin 1 (ET-1), von Willebrand factor (vWF) and interleukin 6 (IL-6), with distinct capillaroscopic SSc patterns and with more severe disease.

Methods: Forty consecutive SSc patients attending our EUSTAR SSc clinic, aged [median (IQR)] 52 (18) years, male gender 4/40 (10%), diffuse cutaneous subset (dcSSc) 14/40 (35%) were enrolled in this study. Extensive clinical and nailfold capillaroscopy (NFC) pattern assessment, as well as quantification of serum E-sel, ICAM-1, ET-1, vWF, IL-6 and C-reactive protein (CRP) were performed on all patients. Associations between vascular biomarkers and disease characteristics were evaluated by Mann-Whitney U-test and Spearman correlations.

Results: NFC "late" pattern was found in 21 patients, while 6 had "early" and 13 had "active" NFC pattern. All 5 vascular biomarkers correlated with each other good to moderately, with r indices varying between 0.660 and 0.332, the only exception being ET-1 which did not correlate with E-sel. Good correlations (r 0.465 to 0.727) were also found between all 5 biomarkers and CRP. Patients with severe vasculopathy, as reflected by the NFC "late" pattern, had higher levels of IL-6 (median 12.06 vs. 3.08 pg/mL, $p=0.001$), ET-1 (median 2.06 vs 1.59 pg/mL, $p=0.029$), vWF (median 3284 vs 2730 IU/mL, $p=0.013$) and E-sel (median 52.6 vs. 42.3 ng/mL, $p>0.05$), compared to patients with NFC "early" or "active" patterns. There was a significant, negative correlation between lung transfer for carbon monoxide (DLCO) and E-sel, ICAM-1 (both $p<0.001$) and vWF ($p=0.013$). ET-1 was higher in patients with more severe disease (dcSSc, patients positive for anti-topoisomerase antibodies and patients with a history of digital ulcers – all $p<0.05$).

Conclusions: Serum biomarkers reflecting endothelial cell activation and/or dysfunction are elevated in patients with more severe SSc-associated vasculopathy and correlate with serum CRP. Together with NFC data they might be used for assessing vasculopathy severity in SSc and identifying patients who would benefit from more aggressive vasoactive treatment.

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FRI0369 PROSPECTIVE EVALUATION OF THE CAPILLAROSCOPIC SKIN ULCER INDEX (CSURI) IN CLINICAL PRACTICE

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Background: Nailfold videocapillaroscopy (NVC) is an imaging technique representing a reliable tool for the classification, diagnosis and monitoring of systemic sclerosis (SSc) patients. The capillaroscopic skin ulcer index (CSURI) was suggested to identify patients at risk of developing digital ulcers (DU) [1].

Objectives: This study aims (1) to describe the practicality of the CSURI in clinical practice, (2) to describe the change of CSURI during follow-up, and (3) to assess associations between the change in CSURI and demographic and disease characteristics.