expenditures were related to higher bDMARD uptake (Table), though not meeting statistical significance (OR 1.91; 95% CI 0.93,3.92). Similar findings were found with country GDP (OR 1.72;95% CI 0.83,3.57).
Conclusions: There remains important residual variation across countries in bDMARD uptake of patients with SpA followed in specialized SpA centers. This is despite adjustment of well-known factors for bDMARD use such as clinical and country-level socio-economic factors.
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Early diagnosis of systemic sclerosis and myositis: biomarkers and diagnostic tool

OP0031 DEVELOPMENT OF A NOVEL EPITOPE-BASED DIAGNOSTIC ASSAY FOR SYSTEMIC SCLEROSIS

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Background: We described the conformational PDGFRα epitope of V_HPAM–Vκ16F4 agonistic autoantibody[1], cloned from memory B cells of a SSc patient, that can induce fibrosis in vivo[2]. We showed that peptides composing this epitope may be specifically recognized by serum IgG of patients with systemic sclerosis (SSc), but not of controls.
Objectives: i. To identify the immunodominant peptide within the discontinuous PDGFRα epitope of V_HPAM–Vκ16F4; ii. to identify other immunodominant epitopes recognized by agonistic autoantibodies; iii. to use these immunodominant peptides to develop an epitope-based assay for diagnosis of SSc and classification of SSc clinical subtypes.
Methods: i.The large PDGFRα peptide library used for epitope mapping of monoclonal anti-PDGFRα antibodies[1] was screened with 25 SSc (12 limited, 13 diffuse) and 25 healthy control (HC) serum samples. ii. A smaller PDGFRα peptide library containing only the top 20 conformational binders plus 20 linear and 20 conformational controls was synthesized. 60 conformational and linear peptides of a cognate protein forming a molecular complex with PDGFRα were included in the array. 20 scrambled peptides were added as negative controls. This library was screened with the same 50 serum samples. iii. A third library was synthesized, retaining the top cognate protein peptide binders, and 15 chimeric PDGFRα/cognate protein peptides, chosen among the best binders, with some nonbinding controls. This library was tested as before. Libraries were synthesized by Pepscan Presto, Netherlands. Statistical analysis was performed by Wilcoxon-Mann-Whitney test. Correlations between serological results and clinical status were made.
Results: i. An immunodominant peptide discriminating SSc from HC serum samples was identified in the first library. ii. This was confirmed by the second library, which highlighted also one immunodominant epitope from the cognate protein. Statistical analysis identified two cohorts of SSc samples (reactive vs nonreactive, the latter undistinguishable from HC) each composed by limited and diffuse SSc subtypes. iii. The third peptide library identified the chimeric peptide recognized exclusively by the reactive SSc serum samples, which were taken from patients with active, progressive disease regardless of limited vs diffuse classification, whereas the nonreactive SSc samples were taken from subjects with less active, non progressive disease.
Conclusions: We developed a conformational epitope-based assay detecting SSc-specific, agonistic, serum autoantibodies. The preliminary results suggest that this novel array may identify SSc patients with active disease, regardless of the canonical classification criteria. We propose this assay for prospective screening of large cohorts of patients affected by, or suspected for, SSc, to validate it as a tool for disease activity assessment and/or early diagnosis.
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OP0032 IS IMMUNOHISTOCHEMISTRY USEFUL TO PREDICT RESPONSE TO TREATMENT IN NECROTIZING MYOPATHIES?

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Background: Muscle biopsy is the gold standard for the diagnosis of inflammatory myopathies, but the role of immunohistochemistry in Necrotizing Myopathies (NM) has not been fully characterized yet.
Objectives: To determine if MHC-I expression and pattern of C5b-9 deposition in capillaries correlate with clinical phenotype and response to treatment in NM.
Methods: The Neuropathology Departmental database was searched to identify patients with a histological diagnosis of NM and follow up data for at least 6 months (30 patients). Electronic patient records were reviewed retrospectively to record demographics, autoantibodies, treatment, proximal muscle power at 3, 6 and 12 months by Manual Muscle Testing (MMT) (2), levels of CK and flares. Patients were classified as responders when there was improvement of MMT $\geq$20% and non-responders when MMT improvement was $<$20% (3). All biopsies were reviewed blindly by an experienced neuropathologist. MHC-I expression was classified as positive only if over expressed in all fibers. The patterns of C5b-9 deposition in endomysial capillaries were classified as specific (solid), non-specific (granular) or negative.
Results: MHC-I positive group (n=16/30) had a higher proportion of responders (62,5% vs 7,7%, p=0.002), higher number of patients with total recovery of muscle power (66,7 vs. 15,4%) and were more commonly positive for autoantibodies (75% vs. 35.7%, p=0.030) when compared to the MHC-I negative group (n=14). 17 patients were positive for auto-antibodies of which 9 were myositis specific antibodies [SRP (n=6), HMG-CoA reductase (n=1), Jo-1 (n=1), P155/140 (n=1)] and 4 were myositis associated antibodies [Ro-52 (n=2), Ku (n=1), Pm/Scl (n=1)]. 13/30 patients had C5b-9 deposition, with a specific pattern in 5 and non-specific in 8. The specific pattern group had a greater reduction of CK after 6 months compared to non-specific and negative respectively (98% vs. 77% vs. 56.8%, p=0.006), greater reduction in CK after 12 months (96.6% vs. 68.9% vs. 59.6%, p=0.024) and higher rates of responders (80% vs. 60% vs. 18,8%, p=0.001). Six patients were on immunosuppressants (azathioprine/hydroxychloroquine, n=2), steroids (n=3) or both (n=1) for a minimum of 4 weeks when the biopsy was performed. Differences in age, gender, clinical features or treatment were not found to be statistically significant.
Conclusions: Upregulation of MHC I and solid staining pattern of C5b-9 in the capillaries of NM patients appears to be associated with a better outcome.
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OP0033 SPECT- AND PET/CT IMAGING IN NEWLY ONSET IDIOPATHIC INFLAMMATORY MYOPATHY

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Background: Diagnosis of idiopathic inflammatory myopathies (IIMs) is challenging and so far no pathognomonic signs exist by imaging. Few radionuclide imaging techniques have been tested for this purpose, mainly ^{99m}Tc-pyrophosphate planar imaging and ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography ($^{18F-}$FDG-PET/CT). However, ^{99m}Tc-PYP uptake has been assessed visually and at best graded semi-quantitatively.
Objectives: We aimed for quantitative ^{99m}Tc-pyrophosphate single photon emission computed tomography/computed tomography (^{99m}Tc-PYP-SPECT/CT) as well as ^{18F}-FDG-PET/CT imaging in a group of newly onset IIM patients.
Methods: Thirteen patients (mean age 62 years) with newly diagnosed, untreated IIM underwent ^{99m}Tc-PYP SPECT/CT of the thorax, pelvis, and thighs. Seven of the patients also had a whole-body ^{18}F-FDG PET/CT scan. Forty-nine healthy controls (mean age 59 years) underwent ^{99m}Tc-PYP SPECT/CT and 26 healthy controls (mean age 57 years) had a ^{18}F-FDG PET/CT scan done. Volumes of interest (VOIs) covering the right biceps, triceps, and quadriceps muscles were drawn manually on each series. Registered ^{99m}Tc-PYP counts, respectively standardized uptake values (SUVs) of ^{18}F-FDG were obtained from all VOIs. Registered counts were decay- and attenuation-corrected and adjusted for body weight and administered dose of ^{99m}Tc-PYP, yielding a parameter similar to the SUV [g mL^{-1}].

Results: IIM patients had visible tracer uptake in the skeletal muscles of the extremities. The muscular ^{99m}Tc-PYP uptake was significantly higher in upper limbs of patients than the uptake in the same muscle groups in healthy controls (uptake[biceps] 0.46 vs. 0.33 g mL^{-1}, p=0.01; uptake[triceps] 0.40 vs. 0.27 g mL^{-1}, p=0.003). The ^{99m}Tc-PYP uptake tended to be higher in the lower limbs of patients than in the lower limbs of controls (uptake[quadriceps] 0.59 vs. 0.48 g mL^{-1}, p=0.06). The muscular FDG uptake was significantly higher in patients than in controls in both upper limbs (SUV_{mean}[biceps] 1.35 vs. 0.72 g mL^{-1}, p=0.006; SUV_{mean}[triceps] 0.91 vs. 0.44 g mL^{-1}, p=0.0008) and lower limbs (SUV_{mean}[quadriceps] 0.84 vs. 0.62 g mL^{-1}, p=0.0001). The muscular FDG uptake values were consequently higher than the ^{99m}Tc-PYP uptake values, although not by a constant factor.
Conclusions: Quantitative ^{99m}Tc-PYP SPECT/CT as well as ^{18}F-FDG PET/CT imaging revealed muscular inflammation in patients with newly onset, untreated IIM. Patients had higher tracer uptake in skeletal muscles groups than healthy controls. Quantification of muscular tracer uptake with the potential to objectively distinguish physiology from pathophysiology could be a valuable tool in the challenging diagnosis of IIMs.
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OP0034 FACTORS ASSOCIATED WITH DISEASE PROGRESSION IN EARLY-DIAGNOSED PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH SYSTEMIC SCLEROSIS: LONGITUDINAL DATA FROM THE DETECT COHORT

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Background: Pulmonary arterial hypertension (PAH) is a severe complication of systemic sclerosis (SSc).
Objectives: In this longitudinal study we aimed to identify factors associated with an unfavourable outcome in SSc patients with early PAH (SSc-PAH) from the DETECT cohort.
Methods: DETECT enrolled patients with SSc fulfilling the 1980 ACR classification criteria, with a minimal disease duration of 3 years since the first non-Raynaud symptom, and with a lung diffusion capacity for CO (DLCO) <60% of the predicted value. A broad range of clinical and laboratory parameters potentially associated with PAH were assessed, and right heart catheterisation (RHC) was performed in all patients at baseline. Patients diagnosed with PAH were followed up for up to 3 years in centers that agreed for the longitudinal part of the DETECT study, collecting data on survival, World Health Organization (WHO) Functional Class (FC), hospitalization, and PAH-specific treatment. Disease progression was defined as the occurrence of any of the following: WHO-FC worsening, PAH therapy with a drug combination, hospitalization, or death. Associations between baseline variables and disease progression were assessed by univariable logistic regression.
Results: Of the 145 SSc patients with PH enrolled in DETECT, 87 patients were diagnosed with PAH, of whom 57 participated in the longitudinal study (median follow-up time 12.6 months, interquartile range 10.7–21.7 months). Among these 57 patients, 33/57 (57.9%) had mild PAH, in WHO FC I or II. During follow-up, 25/57 (43.9%) patients had disease progression (4 deaths, 11 hospitalizations for PAH, 14 with worsening in WHO FC, and 8 received PAH-specific combination treatment), with a 1-year survival rate of 93%. The following factors [odds ratio, (95% confidence interval)] were associated with disease progression: male gender [4.1 (1.2–14.1)], high Forced Vital Capacity (FVC) % predicted/ DLCO % predicted ratio [3.6 (1.2–10.7)], and high Borg dyspnoea index [1.7 (1.1–2.6)]. Low DLCO (% predicted) was also significantly associated with progression [area under the curve (95% CI) 0.8 (0.6–0.9)], but the relationship was not linear.
Conclusions: More than 40% of early-diagnosed SSc-PAH patients in the DETECT cohort who were followed over time had disease progression during a rather short follow-up time, with male gender, functional capacity, and pulmonary function tests (low DLCO, high FVC%/DLCO % predicted ratio) at PAH diagnosis being associated with progression. This suggests that even mild and early detected PAH should be regarded as a high-risk complication of SSc, and every effort to make an early diagnosis is valuable.
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OP0035 PRELIMINARY ANALYSIS OF NAILFOLD CAPILLAROSCOPY IN THE VERY EARLY DIAGNOSIS OF SYSTEMIC SCLEROSIS (VEDOSS): THE CAPI-VEDOSS EXPERIENCE

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Background: In Systemic Sclerosis (SSc), before the onset of clinical signs of fibrosis, puffy fingers, specific autoantibodies and microcirculatory modifications are present and are identified in the VEDOSS criteria for "(very) early" disease. No large scale studies are available indicating the prevalence of nailfold video-capillaroscopic (NVC) SSc patterns or quantitated capillaroscopic characteristics in a "(very) early" cohort.
Objectives: Evaluation of the prevalence of SSc patterns and quantitated capillaroscopic characteristics in a "(very) early" crossectional SSc cohort.
Methods: Multicentre observational cohort study of patients with two strata (Figure): 1.Raynaud's phenomenon (RP), Anti Nuclear Antibody (ANA) positive (+), 2. RP+, ANA negative (-). "Target" (RP+, ANA+, puffy finger [PF] +/-, SSc- antibody (SSc-AB)+, NVC+) and "control" (RP+, ANA-, SSc-AB-, NVC-) subsets were described. NVC patterns (normal/non-specific alterations; non-specific abnormalities; SSc-patterns ("Early", "Active";"Late"; "Sclerodermalike") and quantitative ("absent (="none" or "rare") and "present" (="moderate" or "extensive") capillaroscopic alterations for giants, haemorrhages, capillary loss and abnormally shaped (=bushy capillaries) were evaluated). Generalized Estimating Equations (GEE) were used to assess differences in prevalences of NVC patterns between strata, taking clusters of patients within centers into account.
Results: 1085 RP patients from 40 centres (median number [n°] of pt 12 (minimum [min] 1-maximum [max] 393) per centre were enrolled in the VEDOSS online database. Due to erroneous included/missing data (e.g. erroneous: absence/missing information on RP, presence of former ACR criteria for SSc, skin involvement at baseline; missing info on: ANA positivity, PF, Cap, SSc-AB) 750 patients (median n° of 7 (min:1-max:271) were retained for the analysis. SSc nailfold videocapillaroscopic (NVC) patterns ("Early", "Active", "Late", "Scleroderma like" were present in 79%,13%,0%, 8%) as well as presence of "moderate" or "extensive" giants (49%), hemorrhages (32%), capillary loss (11%) and abnormal shapes (11%) of "target" patients and per definition in 0% of "controls". Estimated distribution of SSc patterns differed in the ANA+/- stratum: 49%/15% ($p \leq 0.001$) ("Early" 40/13%; "Active" 5/2%; "Late" 0/0%; "Scleroderma like" 3/0%). For the quantitative capillaroscopic characteristics a statistically significant difference in the presence of "moderate" or "extensive" giants (23/5%, p=0.027) between the ANA+/- stratum was found, hemorrhages (18/9%, p=0.200), capillary loss (5/2%, p=0.798) and abnormal (ramified) shapes (7/2%,p=0.445).