Abstract OP0145 – Figure 1 Relative change in FVC from base line at different time points

Conclusions: A vast majority of individuals of scleroderma ILD patients on MMF in our cohort had a stable disease or improvement over short and long term followup both in terms of FVC change from baseline as well as HRCT scoring.

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OP0146 IDIOPATHIC INFLAMMATORY MYOPATHIES & INTERSTITIAL LUNG DISEASE

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Background: Idiopathic inflammatory myopathies (IIM) is associated with interstitial lung disease (ILD). IIM associated ILD ranges from subclinical disease, to rapidly progressive ILD (RPILD). Early recognition of these patients is essential for determining treatment.

Objectives: A retrospective case-control study in a tertiary referral centre to identify: a) Clinical features associated with ILD in IIM.

b) Whether antibodies e.g. anti-ENA and myositis specific antibodies (MSA), may aid recognition of ILD or RPILD.

c) Whether intensive immunosuppressants have implication on prognosis of ILD.

Methods: The clinical records of IIM patients who were followed up in rheumatology clinic or admitted into our hospital from Jan 2013 to Dec 2016 were reviewed. We analyse the clinical characteristics (*rash, arthritis, myositis, Raynaud's phenomenon, mechanic hands, and cutaneous ulcers with blood tests*), antibody profile (*anti-ENA: anti-Jo1, Ro, La, Sm, RNP, Scl 70 and MSA: anti-OJ, EJ, PL7, PL12, SRP, PM-Scl75, PM-Scl100, Ku, SAE1, NXP2, TIF1γ, MDA5, Mi2*), treatment and survival. We compare these parameters in IIM-ILD patients against those without ILD. Chi-squared and Mann-Whitney U tests were used to analyse categorical and continuous variables. Log rank test was used to compare survivals.

Results: Among the 101 IIM patients, the mean age was 62 years old with 71% female. 74 patients (73%) had dermatomyositis, 17 (17%) had polymyositis and 10 (10%) clinical amyopathic dermatomyositis. 53 patients (52%) had ILD; 48 (48%) had no ILD. In ILD group, 11/53 patients (21%) were RPILD.

All patients had anti-ENA checked. 59/101 patients (58%) had MSA profile.

Significantly more ILD patients had arthritis, mechanic hands, anti Jo1, anti Ro and anti MDA5 than those without ILD. 21/101 patients had cancers associated with IIM, but cancers were less common in ILD group. Subgroup analyses revealed arthritis, mechanic hands and anti MDA5 were again significantly more common in RPILD compared to other ILD patients (table 1). Anti MDA5 were more commonly found in deceased versus alive patients (40% vs 8.2%, $p=0.02$; OR=7.5). Deceased patients also had significantly higher median peak ferritin (2475 vs 553 pmol/L, $p=0.008$), so did the ILD group (2332 vs 484 pmol/L, $p=0.02$).

ILD patients received more intensive immunosuppressants (high dose steroid, cyclophosphamide, MMF, tacrolimus, IVIg or even rituximab) than non ILD group. The survival was not significantly different between ILD and non ILD groups. However despite intensive immunosuppressants, RPILD patients' survival was still much worse than the other ILD patients (figure 1).

Abstract OP0146 – Table 1 Features with $P<0.05$ and the odds ratios (OR)/relative risks (RR)

	ILD (%)	Non ILD (%)	P value	OR/ RR	RPILD (%)	Other ILD (%)	P value	OR/ RR
Arthritis	53	21	0.001	4.3	73	33	0.02	5.3
Mechanic hands	17	0	0.003	2.1	36	6	0.008	9.7
Carcinoma	8	35	0.001	0.2				
Anti Jo1	20	5	0.03	5				
Anti MDA5	24	0	0.02	2	50	8	0.008	11.8
Anti Ro	40	17	0.01	3.3

Abstract OP0146 – Figure 1 Kaplan Meier curves of RPILD & ILD groups

Conclusions: Certain clinical features and MSA aid recognition of IIM-ILD. Anti MDA5 is related to ILD, RPILD and mortality. Ferritin may be a disease activity and prognostic marker for IIM-ILD. With immunosuppressants, survival of IIM patients with or without ILD is similar. For RPILD patients, the survival is significantly worse despite active treatment.

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OP0147 ABERRANT ACTIVATION OF TYPE I INTERFERON SYSTEM IN ANTI-MDA5 DERMATOMYOSITIS PATIENTS

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Background: Anti-melanoma differentiation-associated gene 5 (MDA5) DM patients have an increased risk of interstitial lung disease (ILD), with a potentially fatal course.¹ Viral infection has been speculated to be the putative trigger for anti-MDA5 DM.^{2,3} The molecular pathogenesis remains largely unknown.

Objectives: In this study, we aimed to explore the role of type I interferon (IFN) system in the pathogenesis of anti-MDA5 DM.

Methods: We studied 20 anti-MDA5 DM patients and compared them with anti-aminoacyl-tRNA synthetase (ARS) DM patients (n=10) and seronegative DM patients (n=30). The levels of IL-1b, IL-4, IL-6, IL-8, IL-10, IL-12, IL-18, TNF-a, IFN-a, IFN-b, IFN-g, B cell activating factor (BAFF), Krebs von den Lungen-6 (KL-6) in blood were tested by enzyme-linked immunosorbent assay and multiplex assay. Expressions of mRNA for sensor molecules (TLR3, TLR4, TLR7, TLR9, MDA5, RIG-1) and type I IFN inducible genes (IRF7, STAT1, MxA, ISG15) in peripheral blood mononuclear cell (PBMC) were detected by real-time polymerase chain reaction analysis. Expressions of STAT1, MxA, ISG15 proteins in skin lesions from anti-MDA5 DM were analysed by immunohistochemistry technique.

Results: Anti-MDA5 DM patients had higher levels of plasma type I IFN (IFN-a, IFN-b), IL-6, IL-10 and TNF-a than seronegative-DM patients. In comparison to anti-ARS DM patients, IFN-a alone displayed heightened level in anti-MDA5 DM patients. Among these 3 subsets of patients, PBMC from anti-MDA5 DM patients

have the significant upregulation of TLR3, TLR7, MDA5, RIG-1 sensors as well as IRF7, STAT1, MxA, ISG15 genes. And skin biopsies from anti-MDA5 DM patients were characterised by strong expression of STAT1, MxA, ISG15 proteins. Furthermore, overexpression of plasma BAFF was observed in anti-MDA5 DM patients. BAFF level was showed to be positively correlated with IFN- α level. Additionally, BAFF level, synergizing with IFN- α , was of great relevance to KL-6 in anti-MDA5 DM patients with higher plasma IFN α concentration.

Conclusions: Our data suggest that aberrant activation of the type I IFN system associated with BAFF may be implicated in the pathogenesis of ILD in anti-MDA5 DM. The discovery may drive the development of new therapeutic strategies for the type of DM patients.

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OP0148

A VALIDATION OF THE 2017 EULAR/ACR IDIOPATHIC INFLAMMATORY MYOPATHIES CLASSIFICATION CRITERIA IN AN EXPERT-DEFINED SINGLE-CENTRE TEN YEAR INCIDENT COHORT

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Background: The recently published 2017 European League Against Rheumatism (EULAR) and American College of Rheumatology (ACR) classification criteria for adult and juvenile idiopathic inflammatory myopathies (IIM) and their major subgroups reflect a long-appreciated need for more accurate case definition in

ongoing research in these complex and heterogenous diseases.¹ However a number of issues remain unresolved. There was a high frequency of missing data in both the derivation and validation samples, only one of the now numerous myositis specific autoantibodies is included, and certain well recognised IIM subtypes are not specifically classified despite their well phenotyped and differing natural histories, clinical features and treatment modalities.^{2,3}

Objectives: To perform an external validation of the EULAR/ACR classification criteria in an incident IIM cohort and examine how classification criteria-assigned IIM subtype correlates with expert opinion.

Methods: Adults with newly diagnosed IIM attending Salford Royal NHS Foundation Trust Neuromuscular services were identified. A retrospective review of all putative cases was performed, and cases fulfilling a consensus expert-opinion diagnosis of definite IIM included. A broad range of clinical, serological and histological data were collected and each case assigned a single IIM subtype by expert opinion. The EULAR/ACR classification criteria were applied and sensitivity, specificity, positive and negative predictive value calculated, presented with 95% confidence intervals (CI).

Results: A total of 922 cases were screened with 255 expert opinion definite IIM identified. The sensitivity to diagnose an IIM was 99.6% (97.2–100) and 80.9% (76.0–85.8) for the classification criteria cut-points of 'probable' and 'definite' respectively. The sensitivity for 'definite' IIM improved to 90.2% (86.5–93.8) when biopsy data for 24/34 initially missed cases were excluded. In 94/255 cases the IIM subtype differed between expert opinion and classification criteria, most strikingly in the group subtyped 'polymyositis' using the EULAR/ACR criteria, where there was discrepancy in the majority (87/161).

Abstract OP0148 – Table 1. PM, polymyositis; DM, dermatomyositis; IBM, inclusion body myositis; ADM, amyopathic dermatomyositis; IMNM, immune-mediated necrotizing myopathy; ASS, anti-synthetase syndrome; OM, overlap myositis.

Classification Tree-assigned IIM Subtype

	PM	DM	IBM	ADM	Totals
PM	37	0	0	0	37
DM	3	54	0	0	57
IBM	0	0	56	0	56
Expert-assigned IIM Subtype					
ADM	0	0	0	14	14
ASS	29	5	0	0	34
IMNM	25	0	0	0	25
OM	30	2	0	0	32
Total	124	61	56	14	255

Conclusions: The criteria performed with high sensitivity in identifying IIM in an external cohort of IIM patients. However, substantial disagreement exists in subtype assignment, especially resulting in a larger proportion of cases of 'polymyositis' with heterogeneous features, important to consider in the application of these criteria to subsequent research.

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