tively after arthritis and inflammatory myopathy onset. Three (12%) patients were admitted to the Intensive Care Unit for Rapidly Progressive (RP) ILD and 2 died (respectively 2 months and 54 months after ILD onset), whereas the alive patient had 2 ICU admission for RP-ILD. Five (20%) patients, including the only 1 dismissed from ICU, needed home O₂ therapy. Ongoing and previous therapies are reported in figure 2.

Conclusion: We showed that the prognosis of IPAF is highly variable, with patients experiencing RP ILD, other slow progressive worsening of respiratory functions and other a substantially stable disease. Furthermore, we showed other findings, laboratory, clinical and instrumental, that could help clinicians in a better identification and stratification of IPAF patients. As a matter of fact, at present, IPAF appears as a generic term including very different conditions that can be further differentiated according to clinical and serological data.

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Figure 1. Antinuclear antibodies determination results

Figure 2. Ongoing (at last follow-up) and previous treatments

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FRI0336 INCIDENCE OF IDIOPATHIC INFLAMMATORY MYOPATHIES (IIM) IN ADULTS IN FIFE, SCOTLAND, UK

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Background:

Objectives: To determine the Adult incidence of IIM in the Kingdom of Fife, Scotland. The Fife population is 370,000 and constitutes 6.8% of the Scottish Population (5,425,000). 98% of the Fife population is Caucasian. We also sought to characterise the incident IIM cases in terms of subtype and autoantibody profile.

Methods: All newly diagnosed Adult cases (≥ 18 years) of IIM between 01.03.17 and 30.11.18 were recorded (11.75 year period). The cases were identified by a search of the Fife Rheumatic Diseases Unit Database using IIM-specific ICD-10 codes. These were cross-referenced with a prospective personal database of IIM cases established by the Lead Author (JMcL) on 26.02.17. In addition a search of all Fife patients enrolled in the UKMYO-NET study was also performed. All patients with 'definite' or 'probable' IIM by the Bohan & Peter 1977 criteria; Clinically defined Inclusion Body Myositis (IBM) by the ENMC 2011 criteria; or other specific criteria in the

case of Clinically Amyopathic Dermatomyositis (CADM) and Immune-mediated Necrotising myopathy (IMNM) were included.

Results: 53 newly diagnosed Adult IIM cases were identified. 11 Dermatomyositis (DM) (3 NXP2; 2 Jo-1; 2 TIF1-G; 1 Mi-2; 3 Myositis-specific antibody (MSA) not tested). 4 Polymyositis (PM) (1 Jo-1; 1 MDA5; 1 MSA/Myositis-associated antibody (MAA) negative; 1 MSA not tested). 12 Anti-synthetase syndrome (8 Jo-1; 3 PL7; 1 PL12) and 3 patients with Jo-1 isolated Interstitial lung disease (ILD). 5 Overlap Connective tissue disease (CTD) : 2 Systemic Lupus Erythematosus; 2 Systemic sclerosis; 1 Mixed CTD. 5 Cancer-associated myositis (CAM) : 2 lung (1 NXP2 DM & 1 SRP PM); 1 ovarian (DM, MSA not tested); 1 Breast (DM, TIF1-G); 1 Prostate (DM, Mi-2). 5 CADM (2 MDA5; 1 TIF1-G; 1 NXP2; 1 MSA not tested). 4 IMNM (2 HMGCR; 1 HMGCR overlap with IBM; 1 SRP overlap with IBM who developed bladder cancer < 3 years after diagnosis). 4 sporadic IBM (2 cN1-A/Mup44 of which 1 overlap with Primary Sjogren's Syndrome; 2 MAA not tested).

Conclusion: The incidence of IIM in Fife, Scotland is 4.5 cases per year which equates to an annual incidence of 12.2 cases per million population. This is comparable with the incidence data published for Salford in the UK (17.6). However the IIM subtypes differed from Salford in that Fife has a much lower incidence of PM. 40/49 (82%) of Fife patients who were tested were found to be MSA positive.

Disclosure of Interests: None declared

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FRI0337 INCIDENCE AND PREVALENCE OF MYOSITIS ASSESSED BY MULTI-SOURCES CAPTURE-RECAPTURE METHODOLOGY

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Background: Precise epidemiology of myositis epidemiology remains largely unknown (1). Surveys based solely on administrative claims benefit from large case ascertainment but may be influenced by miscoding and misdiagnosing. The use of medical records with charts review benefit from accurate diagnosis but exhaustive ascertainment is difficult to achieve because numerous specialists are involved and cases may concern both inpatients and outpatients.

To overcome these difficulties we undertook a capture-recapture survey that takes advantage of a multi-sources case ascertainment to estimate the number of cases missed by any one source and to correct the prevalence rate (2).

Objectives: To assess the incidence and prevalence of myositis in Alsace, a region of eastern France.

Methods: Alsace, region of eastern France, is home to about 2 million inhabitants benefiting from high access to healthcare and a labialized referral center for myositis. Seeking care outside is uneasy because of peculiar geography. Myositis patients were retrieved through three separate sources: i) all general practitioners and community specialist ii) Muscle pathology center records, iii) all public and private hospitals records, vi) all public and private laboratory records. Incident and prevalent cases fulfilling the ACR/EULAR criteria for myositis were included.

Results: The responses to the questionnaires sent to the physicians (n=3452), yielded 105 potential myositis cases. All hospital centres contacted (n=13) participated in the study and 1335 potential myositis patients were recorded by this source. 263 potential myositis cases were identified through muscle pathology center records. 13 laboratories participated in the studies and 324 potential myositis patients were recorded by this source. We thus received 1863 records of suspected myositis after excluding duplicates within each sources. The thorough review of the corresponding medical charts is currently ongoing and at this stage 10% of the potential cases fulfilled the ACR/EULAR criteria for myositis.

Conclusion: This first study based on a multi-sources capture-recapture methodology and ACR/EULAR criteria is very likely to provide an accurate estimation of myositis epidemiology.

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FRI0338 ANTI-MDA5 IDIOPATHIC INFLAMMATORY MYOSITIS (IIM) CONFERS POOR PROGNOSIS BUT NEGATIVE MYOSITIS SPECIFIC ANTIBODY (MSA) IS NOT BENIGN EITHER

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Background: MSA test is useful to diagnose IIM and subcategorize patients by disease phenotypes.

Objectives: The study aims to evaluate the survival of IIM patients of different MSA patterns.

Methods: An IIM registry had been set up in a tertiary referral centre since 2014 by recruiting prevalent and incident cases. Patients were followed-up prospectively. This study included patients fulfilling the 2017 EULAR/ACR classification criteria for IIM¹ and excluded those aged < 18 at disease onset. Immunoblot EUROLINE autoimmune inflammatory myopathies 16 antigens strip (EUROIMMUNE AG, Lubeck, Germany) was used. Information including baseline demographic data, disease manifestations, MSA results, co-existing malignancy, duration of survival and causes of death were collected. IIM patients were divided into seven groups, which included 1) anti-aminoacyl tRNA synthetase (ARS) 2) anti-MDA5, 3) anti-TIF1γ/anti-NXP2, 4) double positive MSA, 5) other MSAs, 6) negative MSA/MAA (myositis associated antibodies) and 7) positive MAA only. Survival probabilities were compared among different MSA groups by using the Kaplan-Meier method and log-rank test. A two-tailed probability value (p) < 0.05 was considered significant. The study was approved by Kowloon Central Cluster Ethic Committee (ref.: KC/KE-17-0177/ER-3)

Results: Among 112 IIM patients, 79 (70.5%) were female, and the median age of onset was 55 (18-90) years old; 63.4% were dermatomyositis (DM), 17.9% polymyositis (PM) and 18.8% clinically amyotrophic DM (CADM). Co-existing interstitial lung disease (ILD) was common and found in 65 (58%) patients; 16 (14.3%) had rapidly progressive interstitial lung disease (RPILD), and 16 (14.3%) died within the observed period. Overall, the commonest cause of death was RPILD, followed by infection and malignancy. While anti-MDA5 was strongly associated with RPILD (odds ratio = 33.0 [95% CI: 7.2-151.8], p<0.001), anti-MDA5 group had the worst survival, with 1-year and 5-year survival both at 43%, compared to above 80% in all other groups (log-rank test p<0.001) (Table 1). There were nine patients with double positive MSA and 28 had negative MSA/MAA. Analysis between MSA sub-groups found that the double positive MSA, MAA only and other MSAs group had no mortality during study period while the negative MSA/MAA group had the second highest mortality following the anti-MDA5 group (Figure 1). Infection and malignancy were the two major causes of death in the MSA/MAA negative group.

Conclusion: Anti-MDA5 associated RPILD was the leading cause of mortality in IIM. However, those tested negative for both MSA and MAA by current immunoblot technique also had guarded prognosis related to the risk of infection and malignancy.

Figure 1. Kaplan-Meier survival of IIM

Figure 1. Kaplan-Meier survival of IIM

Table 1. Survival rates in different MSA groups

Survival	1 year	5 years
Anti-ARS	100%	92%
Anti-MDA5	43%	43%
Double positive	100%	100%
Negative MSA/MAA	89%	82%
Only positive MAA	100%	100%
Anti-TIF1γ/anti-NXP2	100%	88%
Other MSAs	100%	100%

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FRI0339 CLINICAL CHARACTERISTICS OF ANTI-RO52A AND ANTI-RO52B ANTIBODIES IN DERMATOMYOSITIS/POLYMYOSITIS

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Background: The anti-Ro52 antibody, found in numerous systemic autoimmune conditions, is one of the most common autoantibodies in inflammatory myositis. It is known to be associated with interstitial lung disease (ILD) and to coexist with anti-Jo-1¹. There are two spliced forms: Ro52α and Ro52β. Ro52β was originally reported in 1995 as a spliced form of Ro52α in the human heart². Ro52β antigen cDNA has a defective exon 4, which encodes part of an autoepitope hotspot².

Objectives: We investigated the clinical and laboratory characteristics of anti-Ro52α and anti-Ro52β. We also analyzed the characteristics of anti-Ro52α/β within each coexisting inflammatory myositis specific autoantibody (MSA) group to reduce the influences of coexisting MSA characteristics.

Methods: Among 229 dermatomyositis (DM) and polymyositis (PM) patients, 167 patients (DM 152, PM 10, juvenile DM 5) fulfilled the criteria of Bohan and Peter, and 62 clinical amyopathic DM cases fulfilled the criteria of Sontheimer. Anti-Ro52α and anti-Ro52β antibodies were detected by ELISA.

Results: 46 of the 229 patients were anti-Ro52α-positive (20%). ILD was a frequent complication in the anti-Ro52α-positive patients (35/42: 76%) (P=0.0016), and anti-Ro52α was highly positive in the anti-aminoacyl tRNA synthetase (ARS) antibody-positive group (19/32: 60%) (P<0.0001) (Table 1). However, the ILD frequencies of anti-Ro52α-positive and anti-Ro52α-negative patients within each group of coexisting MSA (anti-ARS, anti-TIF, anti-MDA5, etc.) were almost same (P=1). The anti-Ro52α-positive rates were similar to the positive rates of anti-Jo1 (61%) and other anti-ARSS (52%) (P=0.7).

Of the 46 anti-Ro52α-positive patients, 26 patients were anti-Ro52β-positive. No patient was only anti-Ro52β positive. The anti-Ro52α-positive and anti-Ro52β-positive groups were older than the anti-Ro52α-only-positive group (P=0.009), and the average of maximum creatine kinase was higher in both of the positive groups (P=0.065). The 6 patients without coexisting MSA were all both anti-Ro52α-positive and anti-Ro52β-positive (P=0.03) (Table 2).

Conclusion: Anti-Ro52 is highly positive when anti-ARS antibodies are present, but it is not anti-Jo-1-specific. Anti-Ro52 positivity is not associated with elevated risk of ILD in any of the MSA-positive groups. The anti-Ro52β antibody might be an indicator of myositis.

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