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.

REFERENCES:

Disclosure of Interests: Emiliano Marasco: None declared, Federica Meloni: None declared, Giovanni Zantramundo: None declared, Adele Valen
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cucco Speakers bureau: AbbVie, Bristol-Myers Squibb, Celgene, Sanofi, Genzyme, Lilly, MSD, Pfizer, UCB, Lorenzo Cavagna: None declared

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 misdiagnosis. 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. 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 participate 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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FRI0337  INCIDENCE OF IDIOPATHIC INFLAMMATORY MYOPATHIES (IM) IN ADULTS IN FIFE, SCOTLAND
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Background:
Objectives: To determine the Adult incidence of IM 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 IM cases in terms of subtype and autoantibody profile.

Methods: All newly diagnosed Adult cases (≥ 18 years) of IM 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 IM-specific ICD-10 codes. These were cross-referenced with a prospective personal database of IM cases established by the Lead Author (JMcL) on 26.02.17. In addition a search of all Fife patients enrolled in the UKIMYO-NET study was also performed. All patients with ‘definite’ or ‘probable’ IM 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. 58 newly diagnosed cases were identified. 11 Dermatomyositis (DM) (3 NXP2; 2 Jo-1; 1 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 (MMA) negative; 1 MSA not tested), 12 Anti-synthetase syndrome (8 Jo-1; 3 PL7; 1 PL12) and 3 patients with Jo-1 isolated Intestinal 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 (1 HMGCR; 1 HMGCR overlap with IBM; 1 SRP overlap with IBM who developed bladder cancer < 3 years after diagnosis), 4 sporadic IBM (2 cN1A-Mup44 of which 1 overlap with Primary Sjogren’s Syndrome; 2 MAA not tested).

Conclusion: The incidence of IM 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 IM subtypes differed from Salford in that Fife has a much lower incidence of PM. 40/49 (82%) of Fife patients who were tested found to be MSA positive.

Disclosure of Interests: None declared

Figure 1. Antinuclear antibodies determination results

Figure 2. Ongoing (at last follow-up) and previous treatments
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 IIM and excluded those aged < 18 at disease onset. Immunoblot EUROLINE autoimmune inflammatory myopathies 16 antigens strip (EUROMMUNE 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) antibody-positive group (29/90: 32%) (P=0.7), 2) anti-MDA5, 3) anti-TIF1/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 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.

Table 1. Survival rates in different MSA groups

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<table>
<thead>
<tr>
<th>Survival</th>
<th>1 year</th>
<th>5 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anti-ARS</td>
<td>100%</td>
<td>92%</td>
</tr>
<tr>
<td>Anti-MDA5</td>
<td>43%</td>
<td>43%</td>
</tr>
<tr>
<td>Double positive</td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td>Negative MSA/MAA</td>
<td>89%</td>
<td>82%</td>
</tr>
<tr>
<td>Only positive MAA</td>
<td>100%</td>
<td>100%</td>
</tr>
<tr>
<td>Anti-TIF1/anti-NXP2</td>
<td>100%</td>
<td>88%</td>
</tr>
<tr>
<td>Other MSAs</td>
<td>100%</td>
<td>100%</td>
</tr>
</tbody>
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REFERENCES:


FR0339 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 autoimmunopathy 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-1. There are two spliced forms: Ro52x and Ro52xβ. Ro52xβ was originally reported in 1995 as a spliced form of Ro52x in the human heart2. Ro52xβ antigen cDNA has a defective exon 4, which encodes part of an autoepitope hotspot.

Objectives: We investigated the clinical and laboratory characteristics of anti-Ro52x and anti-Ro52xβ. We also analyzed the characteristics of anti-Ro52xβ 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-Ro52x and anti-Ro52xβ antibodies were detected by ELISA.

Results: 46 of the 229 patients were anti-Ro52x-positive (20%). ILD was a frequent complication in the anti-Ro52x-positive patients (35/42: 76%) (P=0.0016), and anti-Ro52xβ 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-Ro52x-negative patients within each group of coexisting MSA (anti-ARS, anti-TIF, anti-MDA5, etc.) were almost same (P=1). The anti-Ro52xβ-positive rates were similar to the positive rates of anti-Jo1 (61%) and other anti-ARSs (52%) (P=0.7).

Of the 46 anti-Ro52x-positive patients, 26 patients were anti-Ro52xβ-positive. No patient was only anti-Ro52xβ-positive. The anti-Ro52x-positive and anti-Ro52xβ-positive groups were older than the anti-Ro52x-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-Ro52x-positive and anti-Ro52xβ-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.

REFERENCES:
