cohort. Furthermore, predominant CNS or ocular GPA (if PR3/MPO negative) were missed by the new ACR classification criteria. EMA Algorithm missed cases of ANCA vasculitis where histology was not possible or ANCA was positive by IIF.

**Discussion:** The study showed the new criteria for AAV, had good performance in INVAR registry AAV patients. The new criteria had good sensitivity and specificity for classification of GPA, MPA and EGPA compared to EMA algorithm.


**Table 1.** Table showing sensitivity and specificity of the EMA algorithm and the ACR 1990 Criteria compared to the new 2022 classification criteria for AAV in the INVAR cohort

<table>
<thead>
<tr>
<th>GPA</th>
<th>EMA ALGORITHM</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>kappa</th>
</tr>
</thead>
<tbody>
<tr>
<td>ACR 1990</td>
<td>87%</td>
<td>89%</td>
<td>0.332</td>
<td></td>
</tr>
<tr>
<td>MPA</td>
<td>ACR 1990</td>
<td>69%</td>
<td>71%</td>
<td>0.081</td>
</tr>
<tr>
<td>EGPA</td>
<td>ACR 1990</td>
<td>92%</td>
<td>99%</td>
<td>0.711</td>
</tr>
<tr>
<td>EGPA</td>
<td>EMA ALGORITHM</td>
<td>69.2%</td>
<td>94%</td>
<td>0.451</td>
</tr>
</tbody>
</table>

**Figure 1.** Venn Diagram showing classification of AAV in INVAR cohort according to various classification criteria

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**Clinical cases**

**POST183**

**DIAGNOSTIC CHALLENGES IN A PREGNANT PATIENT WITH SLE AND SYSTEMIC SCLEROSIS OVERLAP – PREECLAMPSIA, ACTIVE LUPUS NephRITIS OR RENAL CRISIS?**

**Keywords:** Systemic lupus erythematosus, Pregnancy and reproduction, Systemic sclerosis

**Background:** Pregnancy is an important issue for young women with inflammatory systemic disease and pose a clinical challenge.

**Objectives:** The aim of this case report is to underline the challenges to correctly diagnose pregnancy complications versus disease flare in pregnant women with overlap syndrome.

**Methods:** Clinical information collected from the patient's journal.

**Results:** A 26 years old woman was referred to Karolinska University Hospital because of Raynaud's phenomenon, sclerodactyly, skin rash, palpe ulcerations and recurrent finger ulcerations with necrosis, infections and self-amputation of distal phalanges. Interstitial lung disease was confirmed by high resolution CT and a restrictive pattern on lung function tests. Immunological analyses detected autoantibodies against Scl-70, ribosomal-P, SS-A, SS-B, Ku, as well as high titers of antinuclear antibodies. Complement activation, anemia and lymphopenia were also present. The patient experienced muscle weakness and peripheral muscle MRI and muscle biopsy confirmed myositis. During follow-up she developed pericardial effusion and myocarditis. The patient was diagnosed with overlap syndrome with clinical and serological features of systemic sclerosis, SLE and myositis. The immunosuppressive treatment overlapped the disease course comprised hydroxychloroquine, methotrexate, azathioprine, rituximab and low-dose prednisolone. Later, treatment was switched to mycophenolate mofetil because of flares on the previous regimen. To prevent relapse, she was treated with nifedipine, sildenafil and iloprost infusions. Despite medical advice on pregnancy risks during active disease, the patient stopped medication with mycophenolate at the age of 35 years and became pregnant. Enalapril, spironolactone and sildenafil were discontinued and she was referred to the specialist maternity care. The pregnancy evolved without complications until week 18 when the blood pressure (BP) began to rise.

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**Disclosure of Interests:** None Declared.

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