Background: Aminaphtone (AMI) is a bioflavonoid compound, classically used for "capillary disorders". *In vitro* AMI interferes with adhesion molecules (sELAM-1 and sVCAM-1) and with vascular endothelial cadherin degradation thus defending vessels permeability. Moreover, it counteracts vasoconstriction, downregulating endothelin-1 production at a gene level (1-3). *In vivo* AMI ameliorates clinical symptoms of several clinical conditions, above all Raynaud's phenomenon (RP), either primary or secondary to systemic sclerosis (SSc), as demonstrated by a recent six-month study (4).

Objectives: To evaluate long-term tolerability of standard dosage of AMI in a real-life cohort of SSc patients with secondary RP.

Methods: Seventy-eight SSc patients (mean age 65±13 years; mean disease duration 9±7 years) treated with AMI due to active RP were enrolled (ACR/EULAR 2013 criteria). They were taking various concomitant treatments, including aspirin, calcium-channel blockers, cyclic intravenous iloprost, immunomodulators, endothelin receptor antagonists. SSc patients performed periodic clinical assessments and blood tests on average every four months per clinical practice. Duration of AMI administration, side effects, and self-assessment of Raynaud Condition Score (RCS) in a scale from 0 (absence of pain) to 10 (intolerable pain) were retrospectively taken into account.

Results: Duration of AMI administration was between six and sixty-seven months (mean 31±20 months). AMI was administered at 75 mg *bis in die* dosage, as standard initial posology. At baseline, mean RCS was 7.3±0.8. After 3 months of treatment sixty-four patients (82%) yet referred a subjective improvement of RCS (3.5±0.8, $p=0.03$), whereas 14 patients (18%) were clinically unsatisfied (RCS 6.1±0.4, $p=1.12$). In this last group, posology was increased to 75 mg *tris in die*, with a satisfactory amelioration in further nine patients (93.6%) (RCS 4.0±0.6 $p=0.04$), while five patients (6.4%) definitively discontinued therapy for subjective ineffectiveness within six months. Patients referred a sustained improvement of RCS along the observational period (31±20 months) (last RCS 3.7±0.7, $p=0.03$ vs baseline). During the follow-up, five patients (6.4%) referred headache as side effect: three of them had to reduce AMI posology to 75 mg per day, while maintaining clinical benefits. Periodic blood tests did not reveal any significant alteration attributable to AMI. No other side effects related to the drug appeared during the treatment period.

Conclusion: AMI shows an acceptable medium-long-term tolerability along with sustained efficacy in the management of SSc-related RP, without disabling side effects. However, the retrospective design, the absence of a placebo-control group and the concomitant standard therapy limit the results, and a randomized controlled trial for AMI use in the management of SSc-related RP is desirable.

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AB0579

ANTI-JO1 ANTIBODIES IN A REAL-WORLD POPULATION.

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Background: The presence of anti-aminoacyl transfer RNA synthetase (ARS) autoantibodies is essential for the antisynthetase syndromes (ASSD) diagnosis; as well, Anti-Jo1 ARS positivity is the highest weighted criterion in the 2017 EULAR/ACR criteria for idiopathic inflammatory myopathies (IIM). Previous studies have shown differences in ARS specificity between different blot assays. Nevertheless, an adequate clinical correlation and its detection by another monospecific-assays or indirect immunofluorescence assay (IIFA), can safeguard it.

Objectives: To evaluate the prevalence of Anti-Jo1 ARS and their performance in the ASSD and IIM diagnosis criteria fulfillment in a real-world population.

Methods: We performed an observational retrospective study in one center in which IIFA and a blot assay (EUROLINE ANA Profile 3-IgG that include Anti-Jo1 but not other ARS) were performed in all cases with autoimmune disease suspicion (03/2007-07/2019).

We assessed: 1) Anti-Jo1 ARS prevalence, 2) The rate of Anti-Jo1 positivity in the performed blot assays, 3) The rate of patients with Anti-Jo1 ARS meeting ASSD or IIM criteria, and 4) The rate of true and false positive Anti-Jo1 ARS considering the IIFA and clinical correlation.

Results: A total of 419.361 inhabitants are under the center coverage area at the date, a total of 12.711 blot assays were performed during the observation period, and 61 cases presented Anti-Jo1 ARS positivity. The Anti-Jo1 ARS prevalence in the whole studied population was 0.00014, representing 0.04% positivity of the performed blot assays.

Of those with Anti-Jo1 positivity:

- Only 4 patients (6.6%) met Solomon's ASSD criteria and 26 (42.6%) met Connors ASSD criteria (less strict), representing the 0.0009% and 0.006% of the whole population respectively.
- Five cases (8.2%) presented a possible or probable IIM by Bohan and Peter criteria and 55 cases (90.16%) presented a probable or definitive IIM by EULAR/ACR criteria, representing 0.001% and the 0.013% of the whole population respectively.
- The Anti-Jo1 positivity was not confirmed by a monospecific-assay. Nevertheless, 52 cases (85.25%) presented positive IIFA (>1/80): 27 (51.92%) nuclear, 12 (23.08%) cytoplasmic and 12 (25%) with both patterns. A total of 23 cases presented a fine speckled AC-19/AC-20 pattern; representing the 40.98% of all Anti-Jo1 ARS cases.
- Only 4 cases (6.56%) did not meet any of the ASSD or IIM classification criteria. And only 4 cases (6.56%) fulfilled Solomon ASSD criteria or presented a probable IIM by Bohan and Peter criteria, or a definite IIM by EULAR/ACR criteria. Thus, we can estimate that 6.56% of the cases were clinically false positives and other 6.56% were clinically true positive; leading a gap of 86.88% of cases with Anti-Jo1 ARS that only fulfill Connors ASSD criteria (less strict) or a not complete score to confirm the IIM diagnosis by Bohan and Peter or EULAR/ACR criteria.

Conclusion: The low prevalence of Anti-Jo1 ARS positivity observed and the low number of cases with confirmed ASSD or IIM in the Anti-Jo1 positive detected cases, suggest that:

- It is not efficient to test it by screening.
- It should be tested only under adequate clinical suspicion.
- Probably it is not convenient to include it in not myositis specific blot assays. Despite it is desirable to incorporate the evaluation of myositis specific antibodies in the IIM classification criteria; the differences observed between Bohan and Peter and 2017 EULAR/ACR criteria fulfillment in our series, suggest that the latter could be overweighting the Anti-Jo1 ARS positivity.

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AB0580

GENDER DIFFERENCES IN SYSTEMIC SCLEROSIS-IMPACT ON DISEASE PHENOTYPE AND PROGNOSIS

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Background: The low overall prevalence of systemic sclerosis (SSc) and the low proportion of male patients have resulted in a scarcity of studies assessing sex differences in SSc patients, and contradictory results.

Objectives: To evaluate sex influence on disease characteristics at baseline and then to estimate the effects of sex on disease progression and survival.

Methods: We performed a retrospective observational study using data extract from the EULAR scleroderma trials and research (EUSTAR) cohort 096. 173 patients were analysed (26 males).

The severity of organ system involvement was defined as described previously (1).